BRAIN.

PART I., 1898.

Original Articles and Clinical Cases.

HÆMORRHAGE INTO PONS, SECONDARY LESIONS OF LEMNISCUS, POSTERIOR LONGITUDINAL FASCICULI, AND FLOCCULUS CEREBELLI.

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THE case here reported presents, we think, noteworthy anatomical features in connection with the destination of the lemniscus or fillet, the relations of the posterior longitudinal bundles with the cranial nerve nuclei, and also the probable destination of the two sets of fibres, afferent and efferent, constituting these bundles.

Points of clinical interest also attract attention, such as a hæmorrhage due to diseased arteries in a young woman aged 21, with advanced interstitial changes in the kidneys, and hypertrophy of the left ventricle.

Sarah P., aged 21 years, unmarried. Nothing can be discovered in her family or previous history throwing any light upon the cause of her illness. There was a doubtful notch in one of the upper central incisors.

September 14, 1896.—She was in good health until this day. At 3.50 p.m., while at work, she suddenly called out, and was seen by her companions to be "in a fit." She became very blue, and probably lost consciousness; but no more facts can be obtained. At 7.30 p.m. she was seen at home by some friends who state that she could not speak. At 9 p.m. she spoke, saying the name of one of her bystanding friends. Asked if she were in pain, she put her

hand to her head. The right hand "was constantly twitching;" left hand and arm lay motionless. Vomited about six times through the night. Passed urine naturally. Complained of feeling hot. Slept with both eyes closed; breathing very heavy.

September 15.—Admitted into hospital. Stout, healthy-looking girl. Lies on right side. Temperature normal throughout illness, except on September 16, and September 22, when it rose to 100°.

Understands what is said to her, but what she says is for the most part unintelligible; the defect in speech was deemed to be an imperfect articulation, not aphasia.

Paralyses:—Eyes: Both pupils small, right smaller than left; right reacts to light and accommodation sluggishly; left, promptly. Movements upwards and downwards are good. Some divergent strabismus of right eye on looking upwards. Lateral movements completely paralysed in both eyes; she cannot look to right or to left; nor can she converge at all. Some nystagmus in left eye.

Masticating muscles: See notes next day.

Face: Completely paralysed on right side; cannot close right eye at all.

Soft palate apparently not at all affected.

Tongue protruded pointing to the right.

Arms: Right fairly strong. Left almost powerless; she can just move it, but the movements are uncertain and erratic; no reflexes obtained.

Legs: Left leg rather weak, no more. Both patellar reflexes present.

Sensation: Impaired over whole left side of body.

No physical signs of disease in chest or abdomen. Pulse tension raised; artery distinctly thickened.

At 7.45 p.m. she was found snoring and could not be roused. This had passed off by 8 p.m.

September 16.—Paralyses: Power impaired in right masseter, temporal and internal pterygoid muscles. Articulation still affected.

Sensation: Impaired on both sides of the face, most so on upper part of left side. Both conjunctivæ and sclerotics are anæsthetic. [This probably means, anæsthetic on both sides of the body.] Right cornea is anæsthetic, and there are conjunctivitis and keratitis. Sensation impaired on left side of neck and trunk, and on left arm and leg. No loss of sensation in tongue or palate.

Ophthalmoscopic examination showed nothing unnatural, unless it were slight swelling of the disc, and some blurring of vessels at edge of disc.

Pulse 132, full volume, tension raised, artery thickened. Complains of headache.

Urine 1010 sp. gr., thick cloud of albumen, no sugar. By microscope, triple phosphates, and bladder epithelium. Some tendency to retention of urine, rendering use of catheter necessary now and then.

September 17.—Patient says she remembers her attack; was unconscious a little while. Before the attack she had pins and needles in both hands. When she came to her senses she had no headache.

Paralysis:—Left arm and leg; incomplete in both, but much greater in arm than in leg. Complete in muscles supplied by facial and trigeminal nerves on right side. Tongue deviates to right, but less so than on admission. Articulation defective. Complete in external and internal recti of both eyes. A doubtful weakness of left levator palpebræ. No paralysis of palate. Some tendency to vertical nystagmus in left eye. Swallows well. Respiratory movements natural.

Electrical re-actions normal, except that the right facial muscles do not react to faradic current.

Anæsthesia:—Of the whole left side, excepting a small patch on dorsum pedis. In face, anæsthesia most marked in left upper half of face and scalp. Some impairment of sensibility on right side of face. Complete anæsthesia of right cornea; partial of left. Sensation of tongue, palate and inside of mouth natural. She thinks she hears better with right ear than left, but this is doubtful.

Retention of urine. Urine clouded by a few pus globules.

Temporary swelling of submaxillary salivary glands about this time. Mind pretty clear. Pulse very frequent, 132.

September 18.—Passing urine naturally; much pus; catheter has been used.

September 22.—Tongue protruded straight; both sides react to faradism. An apparent ptosis of left eyelid, which disappears on looking upwards, and is probably due to the ophthalmia.

September 23.—A large patch of superficial keratitis of right cornea and discolouration of iris. Right eyelid swollen and red; much conjunctivitis. Retention of urine has recurred.

September 25.—Mental condition not so good; she cries for her mother in a childish way. Fundus of both eyes normal by ophthalmoscope.

September 28.—Has been gradually getting more and more drowsy the last few days. Lies with mouth open; breathes heavily. Pulse hard, 130. Respirations 24. Temperature subnormal. Eyes as before. Right facial muscles do not respond to the strongest faradic current. Faradic reactions in limbs normal. Some wasting of left arm and leg. Excessive patellar reflex, and ankle clonus on both sides. Sensation not so completely lost on left side as was the case a week ago. Taste seems to be absent on the anterior part of the tongue, normal on posterior part. Says she does not smell so well as formerly. Sight unaffected. Hearing unaffected.

September 30.—Much more drowsy, even falling asleep while being faradised. After this date her pulse and breathing gradually failed; she became unconscious and died on October 3.

The temperature chart shows a rise to 100° F. on the day after admission, and again to 100° on September 22. Between these dates, and after the 22nd, the temperature was normal.

Post-Mortem Examination.

October 4, 1896.—Very thin girl, very pale, teeth good, rigor mortis in legs, not in arms.

Head.—Cranial bones natural. Dr. Gee examined head himself, and dictated following note:

"Longitudinal sinus empty. Convolutions flattened, but no sign of tubercle nor of exudation. No tumour on convexity. The anterior tip of temporo-sphenoidal lobe is adherent. The pons looks a little baggy. On the right middle cerebral artery at its first bifurcation about one inch from its commencement, is an opaque yellow patch surrounding the artery (probably atheroma). No embolus. There is more than the ordinary amount of water in ventricle. Septum lucidum natural. The lyra seems decidedly soft; ? ante-mortem. Plexuses quite natural. The corpora striata and optic thalami natural. Cerebellum externally natural. On opening the valve of Vieussens close behind corpora quadrigemina, and on right side is a reddish-brown protrusion about the size of a cherry stone. The median groove on floor of fourth ventricle is pushed to the left by the tumour. The mass spoken of is above the

auditory striæ. All the nerves at base of brain look natural and are not involved in any effusion. Sylvian aqueduct natural."

Lungs.—Both together 20 oz. Old adhesions at apices. No tubercle.

Heart.—11 oz. Pericardium contained 5 oz. brown turbid fluid. Vessels of pericardium injected. Slight exudation of lymph at upper part of auricles. Tricuspid valve natural, also pulmonary. Mitral valve admitted tips of two fingers, looked thickened, a few vegetations on auricular surface. Aortic valves competent to water test; certainly roughened on ventricular surface, and free edges of lunules eroded in places. Heart muscle pale and rather soft. No fatty striation. Left ventricle somewhat hypertrophied.

Kidneys.—2 oz. each. Very small and granular. Capsule adherent and thickened. Cortex scarcely to be seen. Much fat in hilum. Ureters natural, not dilated. Bladder showed no sign of inflammation, except at urethra, which was probably caused by use of catheter. Some pus in vagina and some injection of its mucous membrane.

Summary.—Granular kidneys. Hypertrophy of left ventricle. Endocarditis, ? acute ulcerative. Acute pericarditis.

General Description of the Hæmorrhage.

This is roughly fusiform, with its longest and narrowest end brainwards. It is situated on the right side, but encroaches a little to the left, having destroyed a considerable length of the median raphe, at the same time pushing aside and dislocating structures immediately to the left of the raphe. The greatest diameters lie at about the junction of the middle with the lower third of the pons (see figs. 1 and 2), and dorsally it bulges into the fourth ventricle, pushing up its floor about a quarter of an inch above the general level, without, however, having burst into the ventricle. Ventrally it erodes in a very irregular manner nearly down to the pyramidal fibres, some of the most dorsal of which may be involved (figs. 11 and 12). Its

spinal or posterior end follows the line of the sixth nucleus, so that except for a very small posterior portion, the whole of the right sixth nucleus may be said to be destroyed. Brainwards the line of hæmorrhage seems to follow and be limited to the right lemniscus or fillet, which nearest the main mass of hæmorrhage is almost totally destroyed. It follows the bundle of fibres, as along a line of least resistance, to a point at about the level of the spinal end of the fourth nucleus.

Method of Examination.

The medulla and pons were hardened for about fourteen days in Müller's fluid. Then slices about $\frac{1}{8}$ in. thick were placed in Marchi's fluid for two to three weeks. These after prolonged washing (sometimes over a week) were imbedded in celloidin, and over 200 sections mounted for examination. Of these about one in three were stained by the Pal method in addition. These were valuable for general anatomical purposes, but for the degeneration the Marchi method alone is far superior.

Detailed Examination of the State of the Surrounding Tissues, Nuclei and Fibres Involved.

Fig. 12 is an outline diagram of a section made a little posterior to the greatest diameter of the hæmorrhage. The hæmorrhage here presents a sinuous outline, with an irregular cavity and a number of smaller fissures due probably to shrinkage under the hardening reagents. The edge is sharply marked by what is evidently recent hæmorrhage for a thickness of from $\frac{1}{16}$ to $\frac{1}{8}$ in. The central mass is of a lighter colour, probably consisting of altered fibrin. It contains a quantity of osmic acid staining fatty *débris*, and also numbers of corpuscles of the size of white corpuscles containing in their substance many black granules of fat. In addition to these are many corpuscular masses, as large as a medium sized ganglion cell loaded with black granules,

and presenting in some cases an indistinct clearer space in the centre suggesting a nucleus. They are probably of the nature of phagocytes loaded with degenerated myeline.

Though the arterial walls must have been much damaged no definite aneurysmal dilatations can be seen anywhere. There are, however, several small independent hæmorrhages in neighbourhood of the main lesion on both sides and in the middle line. (Figs. 13 and 14, *h*.)

Nuclei, Fibres and Nerve Trunks directly Destroyed by the Hæmorrhage. (Primary Lesions.)

Except for a small posterior portion (fig. 16), the whole of the right sixth nucleus is destroyed (figs. 12-15, inclusive). With it, as might be expected, the ascending loop of the seventh is involved, and also the posterior longitudinal bundle. The left sixth nucleus is only slightly if at all affected, and probably only by pressure. The posterior longitudinal bundle on the left side is distinctly degenerated, so presumably some of its fibres must have been injured, but not so completely as on the right side (fig. 12, *lbr*). The hæmorrhage at one point encroaches so much over the middle line as to cut across some of the root fibres of the left sixth nerve (fig. 13). The right lemniscus or fillet is completely involved in destruction. The left cannot be seen to be encroached upon, but it must be so to a certain extent, for there are unmistakable degenerated fibres in it and the lateral fillet above. The transverse fibres of the formatio reticularis (figs. 11, 12, 13, *tr*) are extensively destroyed on the right side, and there is degeneration of them on the left.

The cells of the right motor fifth nucleus are difficult to find, being involved in the hæmorrhage, and such as are found are undergoing fatty changes. The sensory nucleus is also much affected, and in places completely destroyed. The ascending (so called) root of the fifth on the right side, all the way down (figs. 13-21, *Va*), presents secondary degeneration of its ventral third. A lesion, therefore, of the root fibres of the sensory fifth must have existed though such cannot be definitely shown.

SECONDARY DEGENERATION OF CRANIAL NERVES.

Fifth. The *ascending root* on the right side shows the degeneration referred to in the last paragraph (figs. 13-21, *Va*).

The *motor root* fibres seem to be completely degenerated (fig. 11, *Vm*).

Sixth. The root fibres on the right side are absolutely degenerated, as might be expected from the completeness of the primary lesion of the nucleus (figs. 13, 14, 15, *VI*). The most anterior fibres of the left sixth root happening to be involved in hæmorrhage encroaching over the middle line are degenerated (fig. 13). The lesion is very slight, however, only to be shown in a few sections.

Seventh. The position of the hæmorrhage is such that, though the seventh nucleus (figs. 14, 15, *VIIn*) is not directly involved, yet the bundle or loop formed by its fibres, and lying dorso internal to the sixth nucleus, is completely destroyed on the right side. Moreover, the supposed connection between the loop and the third nucleus by way of the posterior longitudinal bundle[1] is also severed. Hence an absolute degeneration of the seventh trunk (figs. 14, 15, *VII*).

The condition of the left seventh is interesting. In figs. 14 and 15, *VIIa*, is shown an obvious but not extensive degeneration of the left seventh trunk. This may be explained by a positive lesion of the most mesial fibres of the ascending loop, but it also appears as if degenerated fibres were passing into the loop at all levels from the posterior longitudinal bundle ventral to it, and from the grey matter of the floor of the ventricle dorsal to it, a structure very rich in medullated nerve fibres (this is further discussed below).

Other Cranial Nerves and Nuclei.

The state of the third, fourth, eleventh, and twelfth nuclei nerves will be considered with the posterior longitudinal bundles.

[1] Tooth and Turner, "Study of a Case of Bulbar Paralysis, &c." BRAIN, XIV., 1891, p. 493.

Secondary Degeneration of the Posterior Longitudinal Bundles.

This is a subject of the greatest interest, for it has long been believed on anatomical and clinical grounds that these bundles of fibres serve as paths of communication between the motor cranial nerve nuclei, and that they may also be the channel by which fibres get from a nucleus of one name into a trunk of another.

This fasciculus is completely destroyed for a short distance on the right side, and extensively so but to a less degree on the left; so that there is a well marked secondary degeneration on both sides. The degeneration is also above and below, and therefore this bundle conducts fibres towards and from the brain and in about equal numbers. (?)

(1) *Degeneration below the Lesion (spinewards).*

In figs. 12, 13, 14, and 15 the bundle on the right side is completely destroyed, its situation being marked in fig. 15 by a collection of deeply staining masses of degenerated myeline (*lb*).

In fig. 15 the bundle on the left side presents the familiar triangular shape. The loop of the seventh nerve (*VIIa*), in this and fig. 14, presents a number of degenerated fibres grouped ventrally and dorsally. Their derivation from the posterior longitudinal bundle is suggested above.

In fig. 16, lower down, and now below the level of the hæmorrhage, the posterior longitudinal bundles (*lb*) on either side appear to be taking a ventral course, so that they are now cut almost longitudinally. The lines of degenerated fibres *seem* to pass into the interolivary layer or fillet (see also fig. 17), but that tract of fibres below the lesion may be said to be practically undegenerated.

The sixth nucleus (*VIn*) presents a number of degenerated fibres, many apparently entering the nucleus from the grey matter of the floor of the ventricle dorsal to it.

In fig. 18, at the level of the vagus (*X*) and hypoglossal (*XII*) nuclei, the same mass of degenerated fibres

(*lb*) can be seen lying mesially to the twelfth roots and diverging with them in the direction of the olives. The twelfth nuclei show a considerable number of degenerated fibres, which as in the nuclei above may be derived from the posterior longitudinal bundles. As in the case of the sixth nucleus also, fibres seem to be entering dorsally.

At the level of the post-pyramidal decussation of the lemniscus (*dlm*, fig. 19), which is natural and undegenerated, we still find between it and the twelfth roots the degeneration above described, but here it receives the name of the anterior ground bundle (*ag*).

A little lower at the level of the decussation of the pyramids (*D*, fig. 20), at which level also the anterior horns of the upper cord are now visible, the degenerated fibres are leaving the central parts of the section to be disposed round the anterior horns and on the right side still further appearing in the lateral region to be, as in fig. 21, disposed round the lateral horn, or in other words, the nucleus of the spinal accessory.

We are therefore justified in regarding the anterior ground bundle as consisting largely, if not entirely, of the lower descending fibres of the posterior longitudinal bundle, and as such may be the channel of communication between the spinal accessory nucleus and the higher motor cranial nerve nuclei.

(2) *Degeneration above the Lesion.*

On the right side the degeneration is very complete, less so on the left (figs. 10 and 9, *lb*).

Tracing the bundles brainwards the first interesting connection is with the fourth nucleus (fig. 8, *IVn*), which at this level is practically blended with the posterior longitudinal bundle. Its cells are natural, but the nucleus is pervaded with degenerated fibres, no doubt derived from the posterior longitudinal bundle; in fact, there can be said to be very few normal fibres. The issuing trunk (*IV*, fig. 8) naturally consists largely of normal fibres, but among them appear also several degenerated fibres, apparently passing

away into the trunk without having entered into connection with cells. We infer from this that, as is the case with the motor nuclei hitherto considered, the fourth nucleus receives many fibres from the posterior longitudinal bundles, and that a few of these fibres do not end in the nucleus, but pass straight on into the trunk. May these serve as a connecting link between movements of the superior oblique and external rectus of the same side?

What has been said of the fourth may be equally said of the third nucleus, which is found to be full of degenerated fibres running in every direction. But more than this, we have here an obvious and complete decussation of fibres (fig. 7, 6 and 5, *III*). Among the roots of the third nerves we find a fair number of degenerated fibres (fig. 6), more on the left than the right side. It is tempting to suppose that these are the crossed fibres originating in the sixth nucleus and innervating the opposite internal rectus, for the purpose of conjugate side movements of the eyes, a connection long supposed to exist on clinical grounds.[1]

The crossing of the fibres from one nucleus to the other can be seen to as far as the third nucleus extends (fig. 5). Above this level there still remains a group of degenerated fibres on the mesial side of each red nucleus (*rn*) (figs. 4 and 3, *lb*). These fibres, however, are now taking a direction which causes them to be cut obliquely as if they were passing in a ventral direction. Moreover, there now appears a small bundle of the same fibres (?) in the region of the pillar of the fornix (figs. 4 and 3, *fd*). The destination of these must be conjectural, possibly it may be the corpus mammillare. The fibres of the so-called red nucleus at this level are taking also an oblique course, apparently passing in a lateral direction dorsal to the corpus subthalamicum (*csm*), and among them are many degenerated fibres, possibly derived from the posterior longitudinal bundle. The ultimate destination of many of the fibres may also be the optic thalamus (fig. 3, *ot*).

So that, to sum up, the posterior longitudinal fibres above

[1] Hughes Bennett and Savill, "Case of permanent conjugate deviation . . . lesions limited to sixth nucleus, etc." BRAIN, vol. xiv., p. 102.

the third nucleus seem to send fibres ventrally? to end in the corpus mammillare, laterally with the general trend of the red nucleus fibres, and dorsally to the optic thalamus.

Secondary Degeneration Posterior to the Lesion (Spinewards) of other Fibres.

(1) In the floor of the fourth ventricle there lie normally numbers of fine medullated fibres, which are specially abundant on the dorsal aspect of the various motor nuclei. These fibres, as above noted, are copiously degenerated below the lesion (see figs. 14-18, *d*). It is of course possible that they may constitute an independent commissural system between the nuclei. But this is scarcely probable, and most likely these fibres belong to the posterior longitudinal bundle system.

(2) There is an unmistakable degeneration of the fibres of the antero lateral region shown in figs. 15-19, *at*. This is the situation of the antero lateral ascending tract of Gowers. It was never supposed that this region contained only ascending fibres, but that it resembled the "mixed zone" of the cord. These efferent fibres would seem to be conducted in the formatio reticularis as indicated in figs. 16 and 17, but their actual origin is not known.

(3) A few fibres of the right pyramid are degenerated (figs. 12, 13, *et seq.*, *p*) which can be traced even into the pyramidal decussation (*D*, fig. 20). It is possible that these may be the remains of a considerable pressure lesion at the line of the original hæmorrhage, for no definite solution of continuity of such fibres can be found in any of the sections. That such a pressure did exist is suggested by the transient hemiplegia noted.

Secondary Degeneration of Cerebellar Fibres.

The white matter of the flocculus on either side is extensively degenerated, and in this respect strongly contrasts with that of the rest of the cerebellum (figs. 13-16, *F*). We have been unable to satisfy ourselves as to the origin of the fibres constituting the "arbor vitæ"

of the flocculus, but there is undoubtedly at the level of the maximum lesion an extensive degeneration of the transverse fibres of the formatio reticularis (figs. 11-15, *tr*), and it is obvious that such fibres most probably go to the cerebellum on both sides. They do not, however, appear in the middle peduncle as might be expected, and therefore it is quite likely that they find their way into the flocculus.

Secondary Degeneration of the Lemniscus or Fillet.

The state of this bundle of fibres posterior to the lesion may be disposed of in very few words.

The interolivary layer, the post-pyramidal decussation or decussatio lemnisci, and the funiculi gracilis and cuneatus from which the fillet is believed to arise, are all practically free from degeneration. A few fibres in the interolivary layer are degenerated, but there is reason to believe that they belong to the posterior longitudinal bundles. The fillet, therefore, probably consists of afferent fibres only.

Above the lesion we find that the degeneration of the fillet is complete on the right side, but comparatively slight on the left.

In fig. 11 the main or mesial fillet is still quite replaced by hæmorrhage on the right side. On the left side there is a scattered degeneration in the mesial fillet (*lm*), probably secondary to the encroachment of the hæmorrhage over the middle line shown in fig. 12. On both sides can be seen a marked bundle of degenerated fibres (*ll*) apparently proceeding towards and external to the brachium conjunctivum (*bc*). This is the lateral lemniscus, which anatomically may be regarded as a branch of the mesial, but the fibres of which apparently terminate in the corpus quadrigeminum.

In fig. 10 the hæmorrhage is shrinking to the limits of the right mesial lemniscus. The lateral lemniscus is a marked object in the section; a few degenerated fibres apparently belonging to it seem to cross to the opposite side by way of the lingula (*lg*).

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At the level of the posterior corpus quadrigeminum (fig. 9, *cq*) the fibres of the lateral lemniscus appear to enter and lose themselves in the corpus quadrigeminum. The mesial lemniscus is totally degenerated on the right side, and still contains a small mass of hæmorrhage, much less so on the left. Above the lesion (fig. 7), at the junction of pons with crus cerebri, the same description applies.

In fig. 6 at the level of the third nerve roots the mesial lemniscus alters in the shape of its section, and the fibres apparently become more scattered and so occupy a larger area. Moreover, the disposition of the fibres is such that there is tendency to half surround the red nucleus (*rn*). On the left side, which owing to lateral obliquity of section is more anterior than the right, the fibres of the mesial lemniscus are beginning to take on a lateral direction between the corpus geniculatum internum (*cg*) and the substantia nigra (*sn*).

This lateral direction of the fibres of the mesial lemniscus is still more marked in the section above (fig. 5). A few fibres seem to go to the corpus geniculatum, and the remains of the lateral lemniscus can be seen entering the corpus quadrigeminum.

At the level of the posterior part of the optic thalamus (*ot*, fig. 4) the lemniscus fibres are cut longitudinally, indicating a general change in direction, and suggesting that the greater number of them stream away to the side, that is, away from the optic thalamus, between it and the corpus subthalamicum (*csm*), which here replaces the substantia nigra. This point is also well shown in fig. 3, in which the optic thalamus is well represented on the left side. Unfortunately there was not enough of the brain saved to carry these observations higher or more laterally. It is, therefore, possible that the fibres of the lemniscus may all eventually find their way into the optic thalamus. There can be little doubt that a large number end there, but it is highly probable that many are taking the direction indicated in these diagrams to pursue an uninterrupted course to the cortex.

Remarks.

Reconciliation of Symptoms with Pathological Findings.

The left hemiplegia was no doubt due to pressure on the pyramidal fibres, for the actual damage to the tract is very slight. The hemianæsthesia on the left side might be expected from the very extensive lesion of the right fillet and surrounding structures, formatio reticularis, or tegment (figs. 11, 12 and 13). Mott[1] finds that complete hemianæsthesia does not follow in monkeys after fairly complete degeneration of the lemniscus secondary to lesions of the nuclei funiculi gracilis and cuneatus, but he notes "a distinct lowering of sensibility on the side of the lesion," and that the animal did not use the hand of that side with the same facility as on the other side. So that it would appear that the lemniscus is not the only path of sensory impressions to the brain. This point is confirmed by the experiments of Ferrier and Turner,[2] who regard the tegment as possibly the path of tactile and pain sensations.

The anæsthesia of the upper part of the right face with keratitis follows from the extensive destruction of the right fifth sensory nucleus. The loss of taste on the anterior part of the tongue suggests involvement of the sensory cells of the chorda tympani.

The complete paralysis of the masticatory muscles on the right side might be expected from destruction of the motor nucleus of the fifth, and the consequent secondary degeneration of its root fibres.

The connection between the symptoms and the condition of the sixth nuclei presents certain difficulties. The practically complete destruction of the right sixth nucleus perfectly explains the loss of power of conjugate deviation to the right. But the patient is noted to have had equally complete conjugate paralysis of movement to the left, while the left sixth nucleus is uninjured. The posterior longitu-

[1] Mott, "Experimental Enquiry upon the Afferent Tracts of the Central Nervous System." Brain, vol. xviii., 1895.

[2] Ferrier and Turner, "Cerebro-Cortical Afferent and Efferent Tracts," *Phil. Trans.*, 1897, not published at the time of writing this article.

dinal bundle on the left side is considerably degenerated, but not so much so as on the right, therefore we may assume that the communication between the left sixth nucleus and the right third nerve, or innervation of the right internal rectus, may have been interrupted, but there is still the uninjured sixth nucleus, so that movement of the left eye outwards might have been expected to be unaffected. This was not so, and the facts suggest that conjugate deviation, being a voluntary act, the path connecting the nucleus with the higher centres may be the posterior longitudinal bundle, and that even with a sound sixth nucleus interruption of these fibres may result in such a paralysis. So that it may be regarded as possible that a lesion involving division of the posterior longitudinal bundle on both sides would result in paralysis of conjugate deviation on both sides, even though the sixth nuclei were unaffected.

It is generally believed that the sixth nucleus influences the opposite internal rectus indirectly through the cells of the third nucleus.[1] The present case, however, shows the existence of a certain number of degenerated fibres in the actual roots of the third nerves, which must have been derived from the posterior longitudinal bundles. The discovery of similar degenerated fibres in the fourth nucleus and nerve suggests some association in action between the internal and external recti and the superior oblique.

The situation of the lesion is such that the *seventh* nerve is destroyed at its loop, that is a point at which it would receive the fibres from the third nucleus to innervate the oculo-facial group,[2] and also those supposed to join it from a lower nucleus, ? the twelfth, to supply the lip muscles. As might be expected, the paralysis of the right face was fairly complete, resembling in fact an ordinary facial paralysis. The *nucleus* seems to be intact, but its cells present a large amount of deeply staining fatty granules, more so than on the left side—an example of the "*réaction à distance*" of Marinesco.

[1] Gowers, "Diseases of Nervous System," 1893, second edition, p. 187.
[2] Tooth and Turner, *op. cit.*, p. 490, confirming a former observation by Mendel.

There are undoubted and numerous degenerated fibres in the *left seventh* trunk (fig. 14), which would lead one to expect a certain amount of paresis of the left face. Nothing, however, was noticed wrong with the left facial muscles, but slight weakness of them would have been hard to make out, because the paralysis on the right side would deprive the observer of his standard of comparison.

The eighth nerves and nuclei were unaffected on either side. The double degeneration of the white matter of the flocculi does not appear to have given rise to any symptoms.

A deviation of the tongue to the right, *i.e.*, away from the paralysed side, was noted, but passed off ten days before death. This may have been also a pressure symptom only, the *twelfth* nuclei and pyramidal tracts (except for a very few fibres above mentioned) being practically natural.

Summary.

The following conclusions seem to be warranted on consideration of the foregoing observations.

1. *Lemniscus.*

(*a*) *Constitution.*—Consists of fibres which ascend only, the lowest origin being the nucleus cuneatus and n. gracilis.[1] The lemniscus rapidly increases in size in its progress brainwards, owing, no doubt, to a great accession of fibres from the sensory nuclei of the medulla.

(*b*) *Destination.* — As the corpora quadrigemina are approached the lemniscus gives off a dorsal branch, the lateral lemniscus, which appears to end in the corpora quadrigemina. The main or mesial lemniscus proceeds brainwards towards the optic thalamus, and here again appears to divide into two bundles, one of which loses itself in the optic thalamus; the other mass of fibres appears to take a lateral direction dorsally situated to the corpus subthalamicum. These fibres may enter the optic thalamus higher up, or they may pursue an uninterrupted course to the cortex.

[1] Mott, *op. cit.*, p. 16.

(*c*) *Function.*—Its course and mode of degeneration suggest the transmission of afferent impulses[1] without doubt, but it can scarcely be the only path in the medulla and pons of afferent impressions. Its origin from the terminal nuclei of the two posterior columns of the cord suggest similar functions to those columns.

2. *Posterior Longitudinal Bundles.*

Consist of fibres which run to and from the brain in probably nearly equal proportions.

(*a*) *Descending fibres* appear to serve as a path of communication between the motor nuclei above and below the lesion. They can be traced to the anterior ground bundles of the lower medulla and upper cord, and in fact seem to be the main constituents of these bundles. The position of these fibres suggests a further connection between nuclei above and the anterior and lateral horns of the upper cord, including therefore the nucleus of origin of another cranial nerve, the spinal accessory. A certain homology between the "mixed zone" of the cord and the posterior longitudinal bundle seems probable.

(*b*) *Ascending fibres* enter into close relations with the cells of other motor nuclei (fourth and third). Fibres can also be traced into the roots of the fourth nerve on the same side and into those of the third (? on the opposite side). There is a free decussation across the middle line at the level of the third nucleus. Though most of the posterior longitudinal fibres seem to be absorbed in the third, yet a considerable number can be seen above it, and these probably enter into the optic thalamus.

3. *Flocculus of the Cerebellum.*

Its white matter consists largely of fibres derived from some higher level, probably the transverse fibres of the formatio reticularis.

[1] It must, however, be remembered that Kingdon and Russell describe a degeneration of the fillet apparently secondary to extensive cortical degeneration in children, *Med. Chi. Trans.*, vol. lxxx., 1897, p. 109.

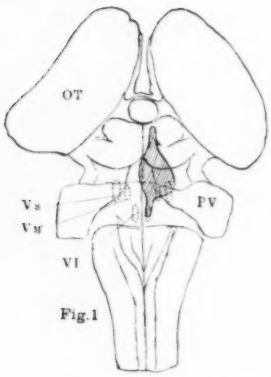
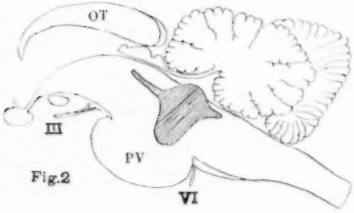


Fig.1



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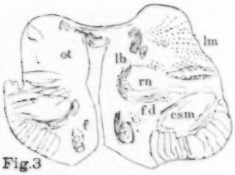

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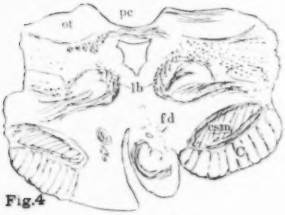

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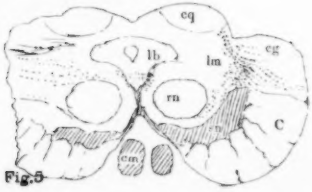

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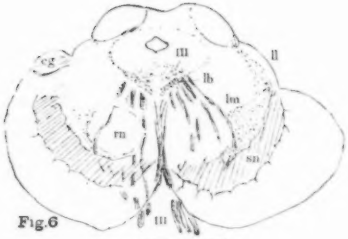
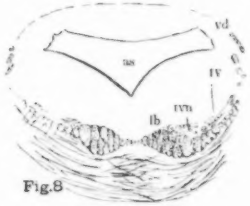


Fig.6

Fig.7



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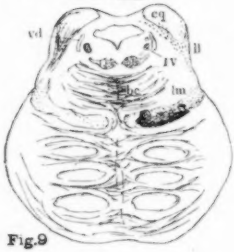

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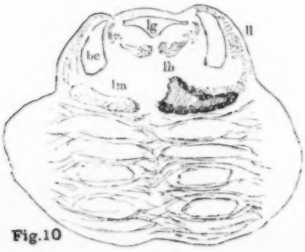

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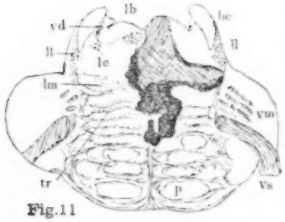

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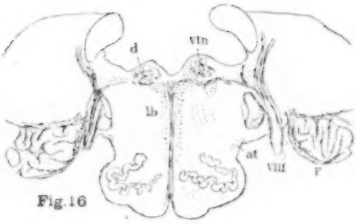

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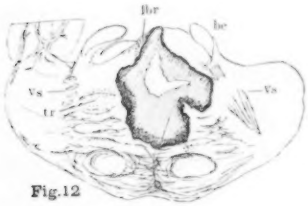

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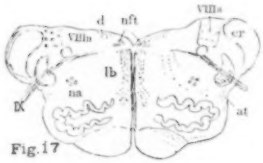
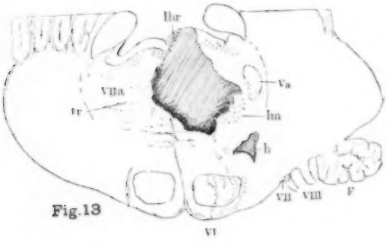


Fig.17



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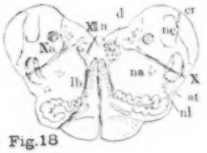

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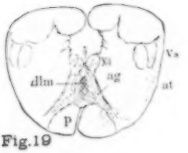

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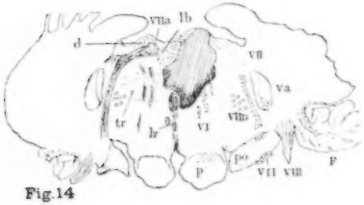

Fig.14

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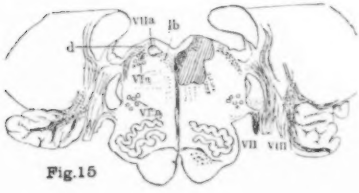

Fig.15

Fig.21

DESCRIPTION OF FIGURES.

FIG. 1.—Hæmorrhage viewed from above. *OT*, optic thalamus; *PV*, pons; *Vs*, sensory, *Vm*, motor nucleus of fifth; *VI*, sixth nucleus.

FIG. 2.—Side view of hæmorrhage.

FIG. 3.—Section (laterally oblique so that left half is at a higher level than right) at about the most posterior part of optic thalamus (*OT*) and third ventricle; *lm*, lemniscus; *f*, pillars of fornix; *fd*, a few degenerated fibres apparently belonging to the posterior longitudinal bundle (*lb*); *rn*, red nucleus; *csm*, corpus subthalamicum with crus ventral to it.

FIG. 4.—A little below the last. *c*, crus cerebri. (Other letters as above.)

FIG. 5.—At highest level of third nucleus on left side; *cq*, corpus quadrigeminum; *cg*, corpus geniculatum internum; *cm*, corpus mammillare; *sn*, substantia nigra.

FIG. 6.—Through the roots of the third nerves. *III*, third nuclei represented by little circles, the dots indicating degenerating fibres decussating; *ll*, lateral lemniscus.

FIG. 7.—Through the lowest part of the third nuclei; *bc*, brachium conjunctivum.

FIG. 8.—Enlarged diagram of central grey tube at level of fourth nucleus (*IVn*) showing its close relation with posterior longitudinal bundle (*lb*) with degenerating fibres in the issuing fourth nerve (*IV*); *as*, aqueduct of Sylvius; *Vd*, descending root of fifth.

FIG. 9.—Through pons at level of the posterior corpora quadrigemina (*cq*). Hæmorrhage on right side lying in totally degenerated lemniscus. Left lemniscus only slightly degenerated.

FIG. 10.—Below the corpora quadrigemina. Lateral lemniscus totally degenerated on right side, less so on left side passing outside the brachium conjunctivum and apparently sending a few fibres across in the lingula (*lg*).

FIG. 11.—At level of fifth nerves. *Vs*, sensory, and *Vm*, motor, about mid pons; motor roots of fifth totally degenerated owing to destruction of nucleus lower down; *lc*, locus cæruleus; *p*, pyramidal tract; *tr*, transverse fibres of substantia reticularis extensively degenerated. The deeply shaded part of the hæmorrhage represents a probably more recent effusion of blood.

FIG. 12.—A little below the last through the largest diameter of the hæmorrhage. *lbr*, right posterior longitudinal bundle totally destroyed. Left considerably degenerated.

FIG. 13.—At the upper level of the ascending loop of the seventh. *Va*, "ascending root" of fifth slightly degenerated in its ventral third; *VI*, right totally degenerated; *h*, small hæmorrhage; *F*, flocculus.

FIG. 14.—At level of seventh and sixth nerves, totally degenerated on the right side. Slightly also seventh derived probably from posterior longitudinal bundle, and from degenerated fibres which lie in the grey matter dorsal to it (*d*). *h*, small hæmorrhages; *VIIa*, nucleus of seventh; *p*, pyramid with a few degenerated fibres in it.

FIG. 15.—At level of upper end of olive.

FIG. 16.—Below hæmorrhage at level of lower part of sixth nuclei (*VIn*). Posterior longitudinal bundle fibres taking a vertical direction; *at*, a degeneration of descending fibres in the antero-lateral tract.

FIG. 17.—At level of ninth nerve. *VIIIn*, eighth nucleus; *VIIIa*, ascending root of eighth; *nft*, nucleus funiculi teretis; *cr*, corpus restiforme; *na*, nucleus ambiguus.

FIG. 18.—At level of upper part of twelfth nucleus (*XIIn*) and nerves, posterior longitudinal bundles lying inside the twelfth roots. *nc*, nucleus caudatus; *nl*, nucleus lateralis; *IXa*, ascending root of ninth.

FIG. 19.—Lower part of twelfth nucleus. Posterior longitudinal bundle continued as anterior ground bundle (*ag*); *dlm*, decussation of lemniscus undegenerated.

FIG. 20.—At decussation of pyramids. *D*, degenerated fibres of anterior ground bundles spreading round the anterior horns into the anterior root zones. Degeneration also of ascending root of fifth (*Va*), as before.

FIG. 21.—About level of first cervical. Anterior ground bundles still contain many degenerated fibres.

A STUDY OF THE LESIONS IN A CASE OF TRAUMA OF THE CERVICAL REGION OF THE SPINAL CORD SIMULATING SYRINGOMYELIA.

BY JAMES HENDRIE LLOYD, A.M., M.D.

Neurologist to the Philadelphia Hospital, Philadelphia.

In 1894 I published a report of two cases of traumatic affections of the cervical region of the spinal cord simulating syringomyelia.[1] In both of these cases there had been severe injury, involving a fracture of the spine in the cervical region, and evidently a wound to the spinal cord. Both patients had survived for a long term of years after the original accidents, and both presented to a marked degree the clinical aspects of syringomyelia. Thus there was progressive muscular atrophy in the shoulders, arms and hands; spastic paraplegia with exaggerated knee jerks, but without persisting paralysis of the bladder and rectum; some trophic lesions, and, finally, the peculiar dissociation symptom, *i.e.*, preservation of tactile sense with abolition of sensibility to heat, cold and pain. These cases were regarded at the time as instances possibly of pachymeningitis cervicalis hypertrophica, although I distinctly stated that I did not intend to commit myself positively to a statement that they were necessarily or even probably examples of an inflammatory process. They simply conformed to the clinical type of syringomyelia, and I was not satisfied in my mind that they were not degenerative or necrotic rather than inflammatory.

Since the report of these cases one of the patients has died, and the autopsy and microscopic examinations are of such great interest that I desire to embody a report of them in another paper. The case has importance from two standpoints. First, with reference to the etiology of syringo-

[1] *Journal of Mental and Nervous Diseases*, June, 1894.

myelia. Second, with reference to the light which it throws upon sensory conduction in the spinal cord. These subjects will be discussed after a description of the case and the sections.

In reporting the clinical history of the case here I shall abstract briefly the report in the former paper just referred to.

Patrick C., aged 60 years, white, male, labourer, had received when 30 years of age an injury to the cervical and dorsal spine by being jammed between two canal boats in a canal lock. For treatment he said that he was kept strapped to a long dorsal splint or platform for five weeks. He must have made a good recovery, for in about three months he returned to work as a common labourer, and continued to do hard work up to the time when he received a second severe accident. This second accident happened about two years before his admission to the hospital, hence after an interval of nearly thirty years. He was in good health at the time. While digging out a clay bank he was buried under a landslide, where he remained about one half hour before he was rescued. He was unconscious for twenty-four hours. On recovering consciousness it was found that he had paralysis in the left arm and leg. The face was not involved and there was no affection of speech. He remained in the hospital for six months. Contractures gradually began in the affected arm and leg.

His condition when first seen by me, three years after the accident, was as follows:

There was an angular deformity, involving the seventh cervical and first and second dorsal vertebræ, and there was a depression also in the region of the lumbar vertebræ. The deformity caused the patient to carry his head in a set position, his chin almost resting on the thorax (fig. 1). The left arm and leg were paralysed, and there was contracture of the arm and forearm. The shoulder muscles and muscles of the arm were paralysed and atrophied, and there were fibrillary contractions in them. Clonus was marked in the fingers, wrist, and ankle of both sides, and the phenomena of so-called spinal epilepsy were readily obtained in the legs. This general leg clonus once started would continue almost indefinitely.

The sensory symptoms were of great interest. The patient was unable to distinguish heat from cold or to recognise either in the region marked on the chart. It is to be observed that this thermo-anæsthesia was on the right side or the side opposite the

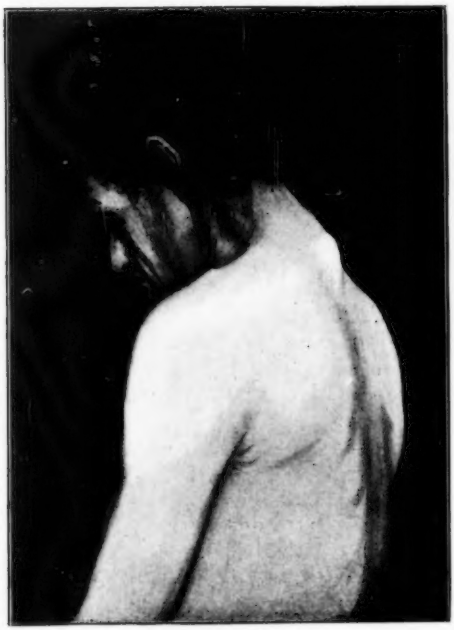

FIG. 1.

A case of trauma of the cervical region of the spinal cord.

paralysis of motion (figs. 2 and 3). There was one area of anæsthesia on the right leg (fig. 4). There was also a form of hyperæsthesia or rather paræsthesia of the right or unparalysed side. Thus he experienced and complained much of a sensation of burning on this side, and wanted constantly to put cold water on it to allay the heat, but he found this remedy ineffectual.

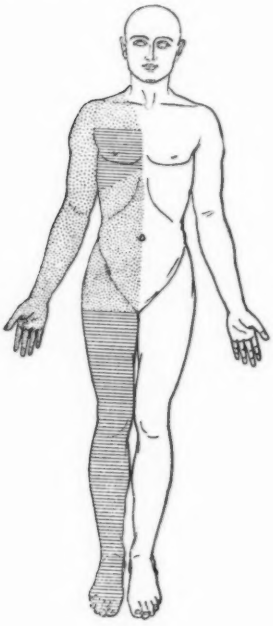

FIG. 2.

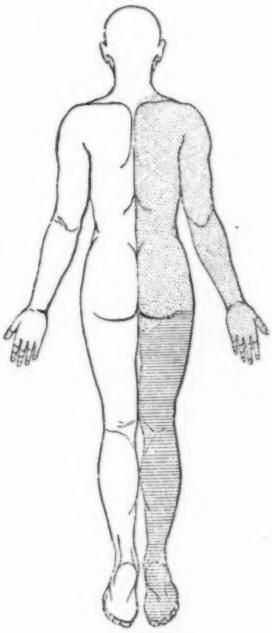

FIG. 3.

Thermo-anæsthesia indicated by cross-lines. Analgesia in the same areas, and also in areas indicated by dotted shading.

Finally, he was profoundly analgesic in the right arm, side and leg. The iris was not involved, nor was there any impairment of respiration or any interference with the innervation of the bladder.

Commenting on this case in the former report I observed that its prominent feature was its hemiplegic type. This

was so pronounced that the case closely resembled one of cerebral lesion. That it was not such, however, was clear from its history, as well as from the deformity of the spine, the absence of paralysis of the face or tongue, the sensory changes and the atrophy and fibrillation in the paralysed muscles. It is to be observed that it followed closely the

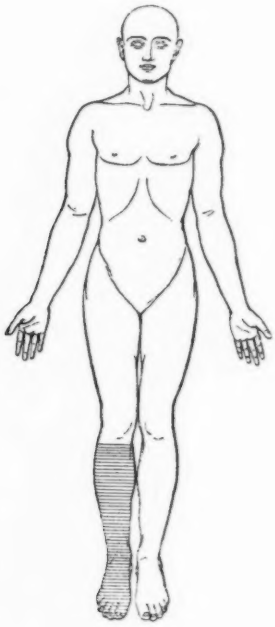

FIG. 4.

Tactile anæsthesia, varied with delayed sensation.

Brown-Sequard type, *i.e.*, the paralysis of motion was on one side and the sensory changes on the other. Its resemblance to syringomyelia was borne out by the peculiar dissociation symptom and the muscular atrophy in the shoulder and arm. As to its hemiplegic type, it must be borne in mind that syringomyelia occasionally presents this

form. That it was the direct effect of accident was evident from the fact that it supervened immediately on the receipt of the injury from the clay bank, which had evidently resulted in a fracture of one or more of the vertebræ of the lower cervical and upper dorsal spine. It seems certain that the injury was the result of the second accident, because the man had completely recovered from the effect of the first accident thirty years before, so that he had been able to do hard work.

The patient died about one year ago from some intercurrent affection. Thus an interval of about five years had elapsed between the receipt of his second injury and his death.

The findings at the autopsy were as follows:

Before the spinal canal was opened, a deformity of the 3rd, 4th, and 5th cervical vertebræ was seen. The spinous processes of these vertebræ were gone, and an osseous mass, resembling callus, was seen on either side of the spinal column. The cord was removed quite easily up to the 6th cervical vertebræ, where the dura was adherent anteriorly to the body of the vertebra, but was free posteriorly. At the junction of the 4th and 5th cervical vertebræ an angular deformity, projecting into the spinal canal, compressed the cord, which was here ribbon-like. The point of greatest compression was at this prominence. The bodies of the 3rd and 4th cervical vertebræ were flattened. Again, at the 2nd and 3rd cervical vertebræ there was an angular projection, caused by a fracture and dislocation of these two vertebræ, with, however, no corresponding deformity in the spinal canal. On the body of the 7th cervical vertebra there was a sharp projecting point. It seemed to be an exostosis, but there was no other evidence of fracture in this region. The cord was much flattened antero-posteriorly at the exits of the 5th, 6th and 7th cervical roots. At and above the exit of the 4th cervical roots the cord was little if any deformed. At the exit of the 8th cervical roots the cord began to assume a normal appearance, and nothing abnormal was noted below this level. The dura at the 3rd and 4th roots was fully twice as thick as normal at the

posterior part, but anteriorly it was normal. There was no other distinct evidence of meningitis, unless it was at the 5th, 6th and 7th posterior roots and on the left side only, where the arachnoid was a little more adherent than is usually observed. There was a spiculum of bone attached exactly to the median and lateral parts of the dura at the exit of the 7th roots, and at this point there was a mass of fibrous tissue possibly resulting from external pachymeningitis. In the brain nothing abnormal was noted.

Microscopic Appearances.

Seventh Cervical Segment (fig. 5).—This is the level of greatest destruction. The whole cord is flattened or ribbon-like at this point. The left half of the cord is extensively injured. The area of greatest involvement includes the left antero-lateral column (the pyramidal tract, cerebellar tract, and Gowers' tract), the anterior and posterior horns, the anterior portion of the posterior columns, and the gray and the white commissures. The lesion extends across to the right side of the cord along the commissure, but in its greatest intensity stops before reaching the horns of the right side. The right half of the cord, however, shows some diffused degeneration of slight degree in the antero-lateral column, and the anterior and posterior horns are misshapen and slightly degenerated. It is interesting to note that the lesion occupies much the same ground as most cases of syringomyelia, also that the posterior columns are affected chiefly in their anterior part, *i.e.*, contiguous to the gray commissure. The left half of the cord in the lateral column is a mass of necrosed tissue, including everything except the extreme anterior column and column of Türck, which are well preserved. The affected area of the posterior columns at this level does not include Schultze's comma zones. It is a somewhat irregular mass of degenerated tissue beginning at the gray commissure anteriorly and extending in a posterior direction so as to include not quite one-third of the columns. In a lateral direction it extends

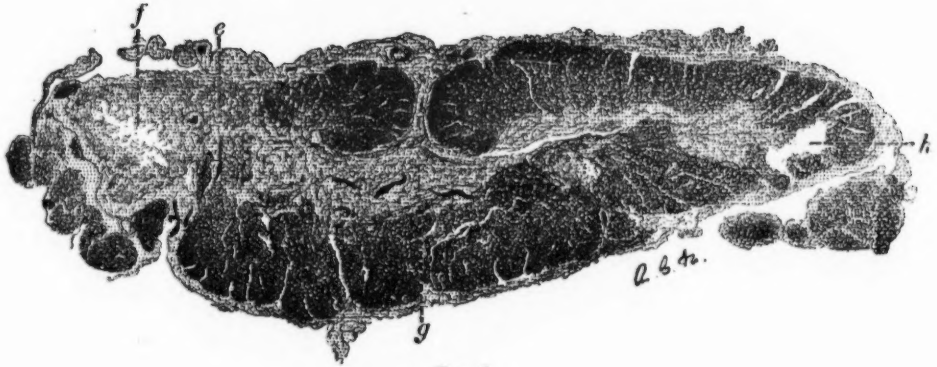

FIG. 5.

Seventh cervical segment.

The section represents the area of greatest degeneration.

(*e*) Degenerated left anterior root.

(*f*) Degenerated lateral column.

(*g*) Degenerated zone about the central canal.

(*h*) Slight area of degeneration in the right lateral column, which cannot be traced very far downward.

irregularly as far as the posterior horns and is rather more extensive in the left posterior column than in the right. It has no definite outline, and cannot be said to be limited to any special group of fibres. It is, however, the starting place for an ascending degeneration in the posterior columns which at higher levels assumes a peculiar branching or W form which will be referred to later.[1] It is thus seen that a large portion of the fibres in the posterior columns escape the lesion (and this doubtless accounts for the preservation of tactile sense).

There is no cavity formation in any part of the cord at this level, nor is any cavity found at any other level of the cord. It is to be noted, however, that the position of the lesion is practically identical with what is seen in many cases of syringomyelia. Thus there is a well marked degenerative zone about the central canal involving both the gray and the white commissures, which at this level are quite obliterated. The only degeneration in the posterior column is that which is contiguous to this central zone of necrosed tissue. This extensive degeneration of the central gray matter of the cord must have acted effectually as a bar to the transmission at this level of any impulses through them from one side to the other. Finally, the extensive necrotic area on the left side, involving both the posterior and anterior horn, the pyramidal tract, the direct cerebellar tract, and Gowers' tract, acts almost though not quite as a hemisection of the cord. The only notable fibres to escape are those in the posterior columns and in the anterior columns and columns of Türck.

Starting from this point we may first trace the ascending degeneration through the higher levels of the cord and,

[1] Bruce ("On the Endogenous or Intrinsic Fibres in the Lumbo-Sacral Cord," BRAIN, part lxxix., 1897), refers to the cornu-commissural tract in the lumbar region, described by Marie as being composed of endogenous fibres. This tract lies in the anterior part of the posterior column in close apposition to the posterior horn, commissure and septum. It does not degenerate in locomotor ataxia, or after injuries to the cauda equina, but degenerates after injuries to the posterior horns, which are supposed to contain its cells of origin. It also degenerates in syringomyelia. This degeneration is said to be descending. It is almost this identical area that has degenerated in my case, only in this instance the tract is in the cervical region, and the degeneration appears to be upward. That these fibres are endogenous is highly probable.

second, the descending degeneration below the seat of the lesion.

Sixth Cervical Segment.—At this level the left half of the cord is also much involved. The degenerated area involves the left pyramidal tract, cerebellar tract and Gowers' tract, and the left posterior and anterior horn. The posterior columns are only involved in the region contiguous to the gray commissure. Hence most of the fibres of the posterior columns escape. The left column is rather more involved than the right. It is to be noted that at this level both the grey and white commissures preserve their normal size, shape, and appearance. There is no dilatation or cavity formation.

Fifth Cervical Segment.—The cord here is still misshapen. The left lateral region is smaller and deeply degenerated. The degeneration in the posterior columns still retains the general outline that was seen at the lower level. The anterior horns are normal in shape and size, but possibly have fewer multipolar cells than normal. The right pyramidal tract and direct cerebellar tract are the seat of a very slight diffuse degeneration. This is seen especially in the pyramidal tract.

Third Cervical Segment.—The posterior columns still present some areas of ascending degeneration. These areas have now taken a more definite shape. They are branching streaks of degeneration passing outward or backward from the region of the gray commissure. The great mass of fibres, however, in the posterior columns are normal. On the left side the cord is still slightly deformed, and here the cerebellar tract and Gowers' tract are deeply degenerated and the crossed pyramidal tract is also the seat of diffuse degeneration. (Ascending degeneration of the pyramidal tract). The central region of the cord (posterior gray and anterior white commissure) is normal. The anterior horns appear at this level to have some degenerated multipolar cells. The right horn especially is smaller than the left, and has not many large cells.

Upper Cervical Cord.—The same general appearances are seen as in the third cervical segment, except that the

anterior horns at this level are normal and contain many large multipolar cells.

Decussation of the Pyramids.—At this level the posterior columns show very plainly two well defined areas of degeneration forming lines or streaks extending first from near the grey commissure in a posterior direction through Burdach's column, and second from a point in Goll's column at the posterior longitudinal fissure backward and outward not extending to the border of the cord. Neither of these zones occupies exactly the region of Schultze's comma zone. It is thus seen, as said already, that the posterior columns are not degenerated in their entirety, but that the vast mass of their fibres escape. It is difficult to account for these peculiarly shaped areas of degeneration in the posterior columns. They maintain the same shape through several segments of the cord above the level of injury. The question may be raised whether these areas of degeneration are caused by the arrangement of the fibres coming in at the point of injury, passing gradually inward toward Goll's column as they ascend. It is possible, however, that they are endogenous fibres, originating in the gray matter at the seat of injury. The cerebellar tract and Gowers' tract are deeply degenerated on the left side.

Medulla Oblongata, Decussation of the Lemniscus.—At this point the direct cerebellar tract and Gowers' tract are still deeply degenerated on the left side. On the right side they are normal. The antero-lateral ground bundles are normal. The nucleus gracilis and nucleus cuniati as compared with sections of normal cords are normal. The posterior columns at this level do not appear degenerated, certainly Burdach's column is not. Goll's columns show deeply stained (non-degenerated) rim of medullated fibres around the nuclei gracilis. (This has been carefully compared with selections from a normal medulla with the result that there may be slight evidence of degeneration in Goll's columns ; if so, this probably represents the group of degenerated fibres running upward through the posterior columns as observed at lower levels.) At this level some of these fibres have probably already terminated in the nuclei

gracilis and cuniati. It is thus of interest to note that at this the highest level of the posterior columns their great mass of fibres is not degenerated. There is no evidence of ascending degeneration of the pyramidal tract beyond one or two segments above the seat of injury. This is in marked contrast with what I found in a case of syringomyelia formerly reported,[1] in which there was deep ascending degeneration of one of the pyramidal tracts extending through and above the decussation of the pyramids.

Descending Degeneration.

Eighth Cervical Segment (fig. 6).—This is just below the seat of injury. The cord is slightly contracted in form on the left side. In the posterior columns Schultze's comma zones are plainly degenerated (descending degeneration). There is also slight diffuse degeneration in the posterior columns which extends only a short distance downward. This is probably caused by the degeneration of the descending branches of the exogenous fibres. It is rather too plainly marked in the illustration. The left lateral pyramidal tract is deeply degenerated, the right only slightly so. The anterior horns contain many multipolar cells some of which are possibly somewhat degenerated. The direct cerebellar tract and Gowers' tract are normal on both sides.

First Dorsal Segment.—The crossed pyramidal tract on the left side is deeply degenerated. This tract on the right side is only slightly degenerated. In the posterior columns Schultze's comma zones are still degenerated. There is also some diffuse degeneration in these columns already referred to (descending branches of exogenous fibres). The anterior horns contain normal multipolar cells.

Second Dorsal Segment. — Deep degeneration of the left lateral pyramidal tract. The slight degeneration in the posterior columns is not so marked as in the preceding section. Schultze's comma zones are not so distinct. Horns normal. From this level downward the cord presents simply deep degeneration in the left lateral pyramidal tract. The posterior columns soon become normal in their entire extent.

[1] *University Medical Magazine*, March, 1893.

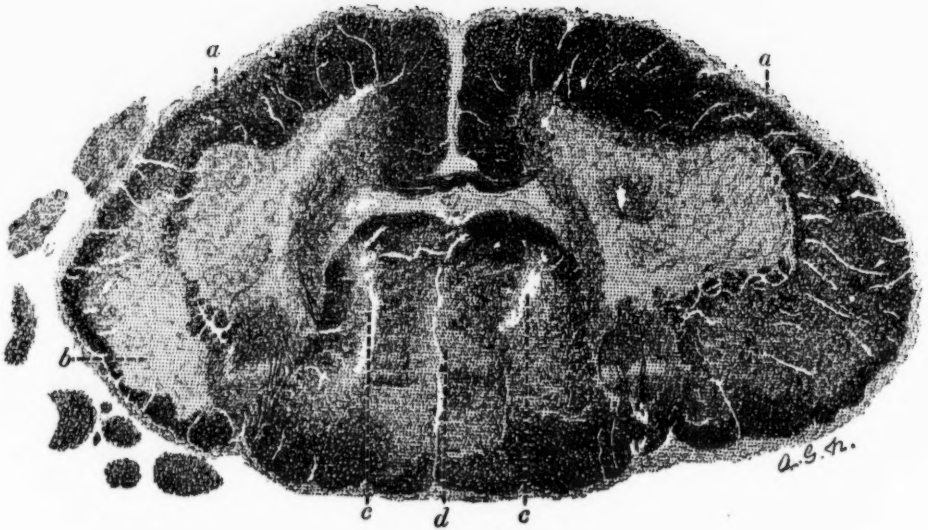

FIG. 6.

Eighth cervical segment.

(*aa*) Anterior horns containing fewer fibres than normal.
(*b*) Degenerated left lateral column.
(*cc*) Degenerated comma zones of Schultze.
(*d*) Slight degeneration of posterior columns which extends only a short distance downwards.

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Comments.

With reference to the etiology of syringomyelia this case may possibly have some significance. It is true that no cavity had formed in this cord, and the case therefore was not one of true syringomyelia. The seat of the lesion, however, especially about the central canal and in the central grey matter, was practically identical with that of many cases of syringomyelia. So true is this that in looking at these sections the observer feels that he is studying a lesion which only required to advance a step further to become truly a case of cavity in the cord. It would seem that it must have been comparatively easy for this necrotic tissue to have broken down still further and to have been absorbed, thus leaving a cavity of irregular extent and outline in the central region of the cord. If this had occurred in the present case the cavity would have occupied especially the region of the grey commissure extending into the posterior column of the left side, and possibly including the lateral column also. For some reason cavities in the cord, as we know, form most readily in this very region of the grey commissure. The causation of them in most cases is obscure, but the possibility of their being caused by trauma has long been recognised. The interesting question arises, therefore, in this case, whether we are not looking at the result of a trauma which has produced an area of necrosed tissue in this usual place without having advanced to the final step of cavity formation. While this question is somewhat speculative the appearances in this cord are highly suggestive of such a state. Cases are not wanting which show the action of trauma in causing cavities in the central grey matter of the spinal cord. Silcock[1] reported a case of fracture of the fifth cervical vertebra, with injury to the spinal cord, in a man who had dived into shallow water. In this case the patient lived for some months. At the autopsy a cavity was found lying immediately inside and parallel to the right posterior horn. There was also much

[1] *Trans. Path. Soc. Lon.*, vol. xxxix., 1888, p. 18.

general sclerosis and disorganisation. Among others who have observed trauma as a cause of syringomyelia are Harckin, Strümpell, and Oppenheim, quoted by Bruhl.[1] They have especially noted falls from high places.

According to Hinsdale[2] the syndrome of syringomyelia is likely to occur suddenly in traumatic cases, and we may then suspect the existence of hæmatomyelia as a starting point for the cavity formation. Hæmorrhages of the spinal cord, as is well known, affect chiefly the central grey matter. According to this same author, Minor observed the symptoms of syringomyelia in organic lesions of the spinal cord resulting from trauma. While it is true that hæmatomyelia may be the starting point for cavity formation in the cord in most cases due to trauma it does not follow that this condition must be the only one. It is supposable at least that necrosis and softening might occur without hæmorrhage and eventually produce a cavity. In the present case there is no evidence of a hæmorrhagic cyst in the cord. All that can be said is that a necrotic area has been produced in the region most favourable for the production of a cavity, and that, clinically, the case had close resemblance to one of syringomyelia. Other cases might be quoted from the extensive literature of this subject.[3]

The question of sensory conduction in the spinal cord is still one of some obscurity. So far as one pathological case can throw light upon this subject some inferences may possibly be drawn from the case under consideration. In the first place is to be noted that tactile sensibility was not involved, except in a limited area on the right lower extremity. Why this comparatively limited and isolated area was involved does not appear plain from the microscopic appearances. It is to be observed, however, that this tactile anæsthesia preserved the shape and extent of the ordinary segmental anæsthesia so commonly seen in hysteria. Thus it involved the whole of the foot and leg as high as the knee where it

[1] "Contribution a l'étude de la Syringomyélie," Bruhl, Paris, 1890.

[2] "Syringomyelia," Alvarenga Prize Essay, College of Physicians of Philadelphia, 1897.

[3] For an extensive bibliography, including more than five hundred references, Hinsdale's Prize Essay, already quoted, may be consulted.

was sharply limited by a line drawn horizontal to the long diameter of the limb. In short, it had the appearance of a stocking, or, as when seen in the arm, of a gauntlet. This segmental anæsthesia, not following the distribution of any particular nerve, is hard to explain by an organic lesion. In considering its significance in this case I have been strongly inclined to regard it as an hysterical phenomenon. The patient had some of the mental characteristics of hysteria, and it may well be that this was one of the hysterical stigmata that not uncommonly complicate the picture of organic nervous disease. That it was not due to organic changes in the posterior columns of the cord is rendered probable by the fact that the columns of Goll at and above the seat of lesion were not seriously involved. Moreover, although slightly implicated, they were equally so on both sides. We know that the sensory fibres from the lower limbs pass upward through the cervical region exclusively in these columns of Goll. Therefore this case presents no reason why total tactile anæsthesia of this peculiar segmental type should be found in only one leg. In the second place, this patient had the peculiar disassociation symptoms of syringomyelia involving the whole right half of the body. The thermo anæsthesia was not so extensive as the analgesia, since the former involved only the lower limbs and a limited region on the thorax. Finally it is to be observed that this hemianæsthesia to pain and temperature was on the side *opposite* to the main lesion in the cord and to the motor paralysis. Hence in a sense it presents the appearance—without tactile anæsthesia, however—of a Brown-Sequard paralysis of the spinal cord.

It seems that it is not difficult in the present case to explain the sensory phenomena by the lesions found under the microscope. In the first place, the preservation of tactile sense is perfectly explained by the fact that the posterior columns of the cord were not seriously involved. The parts of these columns most involved were contiguous to the posterior grey commissure, a region in which the endogenous fibres especially of the posterior columns run. The fibres passing in by the posterior roots (exogenous

fibres) pass up the posterior columns, gradually seeking and entering the columns of Goll. As already said, the columns of Goll are not deeply degenerated at the level of the lesion nor above the level of the lesion. According to Van Gehuchten the probabilities are that these long fibres of the posterior columns, *i.e.*, the exogenous fibres, passing in by the posterior roots, serve for the conduction of tactile sensibility. That is the lesson taught by syringomyelia and also by the present case. These fibres pass up directly into the nuclei graciles and cuniati in the lower part of the medulla oblongata. Here their arborisations are distributed to the neurons of a second order. The microscope in this case does not indicate any marked degeneration of these fibres at or about their point of entrance into these nuclei.

With reference to the thermo anæsthesia and analgesia in this case, I may say that they seem to point unerringly to the conclusion of Van Gehuchten, who says that in all probability pain sense and thermic sense are transmitted through the grey matter by way of cells whose axis cylinders become the fibres which constitute especially the tract of Gowers. These fibres according to the same authority are *crossed*. In the present case Gowers' tract as well as the direct cerebellar tract was deeply degenerated on the *left* side from the seat of lesion all the way up. On the right side there was very slight evidence of degeneration in these two tracts. This would put the case in perfect harmony with the anatomical demonstrations of Van Gehuchten. The tract of Gowers on the left side above the lesion being degenerated and this tract being composed of axis cylinders arising from cells located in all the segments of the grey matter of the opposite side, it follows of course that the thermo anæsthesia and analgesia would be upon the opposite or right side. This was so in this case. The motor symptoms, *i.e.*, spastic paralysis of the arm and leg with muscular atrophy of the shoulder and upper extremity, were most marked in this case, as always, upon the side of the lesion, *i.e.*, the left side. If this view is found to be confirmed by other cases we must accept a modified view of Brown-Sequard's paralysis of the spinal cord. Instead of this

paralysis being characterised by an hemianæsthesia on one side and a hemiplegia on the other we shall have the syndrome constituted as follows:—The motor paralysis and the tactile anæsthesia would be on the side of the lesion, while the anæsthesia to pain and temperature would be on the side opposite the lesion. Of course, as in all pathological cases, the present case is open to the objection that the lesion is not sharply defined nor limited strictly to the one side. Yet in looking without prejudice at the sections it must be acknowledged that the lesion is confined very largely to the left side.

It is but fair to say that all anatomists and physiologists do not as yet agree with Van Gehuchten that the tract of Gowers is composed of crossed fibres, or that it is necessarily the pathway for the pain and temperature sense. Yet the present case points strongly to some such conclusion.

According to Gotch[1] the degenerative changes which follow section of a posterior root are always limited to the side of the lesion and affect only the fibres of the posterior column. This degeneration ceases at the medulla oblongata in the grey matter of the gracile and cuneate nuclei. As the fibres entering the posterior roots divide dichotomously, one branch, already referred to, passing upward, and the other branch looping downward for a short distance to pass into the central grey matter, it follows that in a localised lesion of the posterior columns there might be found some slight descending degeneration in these columns for a short distance below the lesion. There is some evidence of this, but to a very limited extent downward, in this case. Gotch, however, contends that there is no evidence of ascending degeneration on the opposite side of the cord either after a section of a posterior root or after a hemisection. This, however, does not invalidate the claim made here, since the fibres in Gowers' tract and the direct cerebellar tract are *endogenous* fibres, *i.e.*, take their origin in

[1] "Recent Research on the Spinal Cord," by Francis Gotch, M.A., F.R.S., *Liverpool Medical Chi. Journ.*, xiii., 1893, p. 150.

cells located in the grey matter at the various levels of the cord itself. Impulses from painful and thermal impressions, according to this observer, radiate into the grey matter of the cord, and pass particularly into the lateral columns of the opposite side. This is perfectly in accord with our present case. Hence, to recapitulate briefly, it may be said that tactile impressions pass directly up the posterior columns by way of the exogenous fibres of the same side, while painful and thermal impressions pass into the grey matter and through the cell bodies of a second order of neurons whose axis cylinders, in a large majority at least, pass across to the opposite side of the cord and up the lateral columns, especially in Gowers' tract. This seems to be the teaching of the case here under review.

In conclusion, I have to express my obligation to Dr. William G. Spiller for his skilful preparation of the sections in this case, and to Miss Anna G. Newbold, of Philadelphia, for her very accurate drawings of the same.

GENERAL PARALYSIS OF THE INSANE DURING ADOLESCENCE, WITH NOTES OF THREE CASES.

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THE occurrence of general paralysis of the insane in patients about the age of puberty is now an acknowledged fact. Although somewhat uncommon, it is becoming better recognised, and within the last fifteen years a number of cases have been placed on record, gathered chiefly from amongst asylum patients. The three following cases, however, may be of interest, inasmuch as they occurred in the course of ordinary neurological work, and were therefore studied from the point of view of the neurologist rather than that of the alienist. They also help to throw light upon the possible causal antecedents of the disease in patients of so young an age, and they exemplify well its peculiar symptomatology in such patients, whilst one of the cases also presents some interesting points in the study of epileptiform fits.

For permission to publish my notes of these cases I am indebted to the courtesy of Dr. Hughlings Jackson, Dr. Ferrier, Dr. Beevor and Dr. James Taylor.

Case I.—W. H., male, aged $16\frac{11}{12}$ years, was admitted to the National Hospital, Queen Square, on October 1, 1897, under the care of Dr. Hughlings Jackson. He was said to be suffering from "fits."

Family History.—None of patient's relatives have ever suffered from any nervous or mental disease. His father is alive and healthy; his mother died in childbirth at the age of 41. Patient is the second of fifteen children, of whom the first child was born dead and five others died at birth. The remaining members of the family, with the exception of the patient, are perfectly healthy, he being the only delicate member of the family.

Previous Health.—Although never robust, he has not suffered from any definite illnesses beyond the usual ailments of childhood. He never had any convulsions in infancy. There was no history of injury or accident of any sort.

Present Illness.—Patient was formerly a very smart, intelligent boy and very good at his lessons, but about three years ago, when he was 13 years of age, gradual mental dulness came on. The first observed symptom of this was that, when he was sent out on errands, he used to return having completely forgotten several of his messages. He became gradually much duller mentally, quieter and less lively.

About two years ago he had his first "fit." On getting up one morning he complained of feeling sick and went back to bed, when he became unconscious, with pale face and twitching of all his limbs. This twitching is said to have lasted a couple of hours. During the fit he did not bite his tongue, nor were the sphincters affected. After the fit he seemed mentally more peculiar for some days, with hallucinations of vision in the form of moths, &c. These hallucinations passed away in a few days.

Since then he has had three more fits—four fits in all—at intervals of about six months, the last fit occurring six weeks before his admission. For a day or two before each fit he has usually been "quiet and moping." During the fits he has never bitten his tongue, nor has he passed water. In two of the fits he has fallen down at the onset. After each fit for about ten days he has always been mentally peculiar, and his right arm is said to be weak—he "loses the use of it" for a day or two. Patient is naturally right-handed, but now his right hand is said to be much weaker than his left.

Ever since the onset of the illness his father has noticed the patient's gait to be somewhat staggering in character. The boy himself does not complain of giddiness, even when staggering.

Condition on Examination.—Patient is a pale-faced boy of fair muscular development, with sharply cut features. He has a vacant expression and his mouth hangs open. There are no signs of congenital syphilis in the teeth, eyes, face, or elsewhere.

There is considerable mental hebetude. He never volunteers any remark. He answers questions slowly and often incorrectly, speaking in a slow monotonous voice with marked fibrillary trembling of lips, face and eyelids. His memory is very defective. Although usually in a quiet, dreamy mental condition, he has occasional noisy bursts of passion in which he obstinately refuses to comply with simple requests.

Smell, taste, and hearing are normal. The visual fields are not contracted, visual acuity is normal, and the optic discs are healthy.

The pupils are widely dilated, the left more so than the right. They do not react to light, and there is very slight, if any, contraction during convergence. There is no squint, nor nystagmus.

The facial movements are normal save for the fibrillary tremor above noted. The palate rises normally on phonation. The tongue is protruded straight and is not atrophied, but it exhibits marked fibrillary tremors on both sides.

There is no anæsthesia anywhere on the face, trunk, or limbs to the lightest touches or pricks. All the movements of the upper limbs are performed, but in a languid manner, and the grasp of the right hand is slightly weaker than that of the left. In the lower limbs there is a similar languor in all movements, but no actual paralysis. Patient's gait is somewhat ataxic; he walks on a broad base, stamping his heels. He does not sway or fall with feet together and eyes closed. The knee-jerks are brisk and equal. There is no ankle clonus. Supinator and triceps jerks are brisk, and there is marked jaw-jerk.

The plantar, cremasteric and abdominal reflexes are normal and equal on both sides. The sphincters are normal, but owing to his mental dulness he does not always obey the calls of nature, frequently passing both urine and fæces as he walks about. He practises onanism with considerable frequency. The genital organs are much smaller than normal, and there is no pubic hair. The lungs, abdomen and urine are normal. There is a marked systolic murmur at the apex of the heart, conducted outwards to the left side.

The patient remained in the same dreamy mental condition for a month after his admission, with occasional attacks during which he was noisy and passionate. He never expressed exalted ideas of any sort.

On *November* 3rd patient was seized with epileptiform convulsions which came on during the night when he was asleep, and which lasted for nearly four hours without intermission. The fit commenced with a cry, and the twitching was observed to begin in the fingers of the right hand. Owing to the long duration of the fit I had an opportunity of studying its character with some minuteness. The patient, who was completely unconscious, lay on his back, with rapid clonic spasms of all the muscles of the limbs, trunk and face, the right side being distinctly more severely

affected than the left. His face was natural in colour. The eyes were open but with occasional clonic spasms of the orbiculares palpebrarum. There was marked conjugate deviation of the eyes to the right side. The pupils were widely dilated, equal, and insensitive to light. The mouth was widely open. There were no twitchings of the jaw muscles, but the face twitched markedly, especially on the right side. The palate twitched markedly and symmetrically on the two sides. Laryngeal examination showed rapid alternate separation and approximation of the vocal cords, and also alternate narrowing and widening of the aryteno-epiglottidean folds.[1] There was also distinct twitching of the tongue on both sides. All the muscles of the limbs and trunk on both sides twitched briskly and synchronously with the face and tongue muscles. There was marked twitching of all the muscles above and below the hyoid bone, and of both sterno-mastoids equally. The head was drawn backwards and the chin elevated. Respiration was noisy and rendered irregular by the clonic spasms of the respiratory muscles. There was occasional inspiratory stridor. There was no twitching of the anus.

The clonic spasms of all these muscles were irregular in rhythm, the number of contractions counted in a minute being about 180. Patient's skin was warm and moist; the temperature 98·2° F., pulse 156, and the respirations 36 per minute. Patient did not pass urine during the fit, nor did he bite his tongue, owing to the jaws being widely open all the time.

During the fit the knee-jerks were elicited readily.

Next day, *November* 4th, even after large doses of chloral and potassium bromide *per rectum*, although the general convulsions had ceased, twitching still persisted in the left side of the face and tongue and in the left hand; with this there were occasional spasms of tonic rigidity in the trunk and non-twitching limbs. Patient was still unconscious; the eyes were blood-shot and directed upwards and to the right side. The knee-jerks were present and equal, not exaggerated. There was no ankle clonus.

[1] The vocal cords have not hitherto, I believe, been observed during an epileptic fit in the human subject. Analogous observations on the larynx have, however, been made by Risien Russell during absinthe convulsions in monkeys, and by Semon and Horsley in excitation of the cerebral cortex. "We have observed," they say, "that in powerful or long-continued excitation of the laryngeal area, true epilepsy of the vocal cords supervenes, gradually spreading to the neighbouring muscles of the face, neck, head and upper limbs. From this we conclude that the epileptic cry is not, as has been so long believed, the result of a discharge primarily from the medulla oblongata, but rather represents a stage in the cortical excitement popularly termed an 'epileptic fit.'" (Semon and Horsley, "On the Relation of the Larynx to the Motor Nervous System," *Deutsche Medicin. Wochenschr.*, 1890, No. 31, p. 10.)

November 7th. — Patient was somewhat brighter; semi-conscious, but did not speak; became able to swallow again. Occasional twitchings still occurred at intervals in the face alone, sometimes on one side, sometimes on the other.

On *November* 8th patient again became more deeply comatose; twitching entirely ceased. The pupils were dilated and insensitive. Cheyne-Stokes respiration set in, and he died from respiratory failure.

The *post-mortem* examination was made by Dr. Risien Russell, who made the following report :—

Autopsy.—A *post-mortem* examination was made eighteen and a-half hours after death. The skull-cap was natural, as was the dura mater; no undue adhesions existed between these two structures, nor was the dura mater thickened or anywhere adherent to the pia-arachnoid. There was slight but distinct general thickening of the lepto-meninges over the convexity of both cerebral hemispheres, where they presented a milky, opaque appearance. So adherent were the meninges to the surface of the convolutions in some parts that it was impossible to remove them without tearing off the superficial layer of the cortex, while from other parts they were comparatively easily removed without any such damage to the underlying brain. There was a good deal of compensatory œdema of the lepto-meninges. The convolutions of both cerebral hemispheres were considerably atrophied, this was, however, not more pronounced in the frontal lobes than elsewhere; but possibly the shrinkage at the upper part of the central convolutions, more especially the ascending parietal, was a little more marked than in any other part. On palpation the cerebral substance gave rise to a greater sense of resistance than natural, and it was found to be unduly firm when sections were made into the hemisphere. On the whole, the grey and white matter appeared to be reduced in equal proportion, though in parts the atrophy of the former seemed the more pronounced.

No macroscopic changes were detected in the centrum ovale, basal ganglia or internal capsule on either side. The lateral ventricles were somewhat large in comparison to the size of the brain, but they contained clear fluid, and the ependyma showed no granular or other macroscopic change.

Beyond thickening of the pia-arachnoid, chiefly over the upper surface of its middle lobe, the cerebellum presented no abnormal features, nor did examination of the pons and medulla reveal anything unusual. There was no obvious lesion of any of the cranial nerves. The lepto-meninges at the base were not speci-

ally thickened, and there was nothing in the appearances met with to suggest that there had at any time been an acute basal meningitis.

The spinal canal was natural, as were the meninges, but the spinal cord was unduly soft and bulged markedly on section —a condition evidently the result of *post-mortem* change. The thoracic and abdominal viscera were natural, and no appearances were met with about the body, or in connection with any of the organs, which could be regarded as evidence of syphilis.

Microscopic Examination.—The central nervous system was carefully preserved for subsequent microscopic examination, and portions were prepared and stained by various methods, including those of Nissl, Marchi and Pal; in addition to which stains, such as aniline blue-black, hæmatoxylin and eosin, were employed. Portions of cerebral cortex, from the frontal and central convolutions of each hemisphere, thus dealt with, revealed on microscopical examination considerable thickening of the leptomeninges, with marked nuclear proliferation. Large bands of thickened pia extend deeply into the substance of the cortex at intervals, while in other parts the pia is separated from the surface of the brain, and no such thickened bands connect the two. There is marked sclerosis of the cerebral substance, with nuclear proliferation, and the walls of the vessels, both of the pia and in the brain, are greatly thickened. Profound degenerative changes of the neurons of the cortex exist; the cells are seen in all stages of atrophy, some almost beyond recognition, and there is obviously a great scarcity of them. All of the cells present are poorly supplied with chromatic material, and some of them, instead of being shrunken and atrophied, are obviously swollen, and in a stage of more recent degeneration. Similarly, degenerative changes are obvious in the axons, whose continuations can be traced, in many instances, as scattered, recently degenerated fibres, as they course through the corona radiata. No obvious degenerative or sclerotic changes are seen in the cerebellum, such as those that have just been described in connection with the cerebrum.

The most pronounced change met with in the spinal cord is a sclerosis of the postero-internal columns, limited to the cervical region, symmetrical, and occupying chiefly about the dorsal two-thirds of these columns. A very narrow band of sclerosis, on each side of the postero-median septum, extends ventrally from the main area of sclerosis towards the posterior commissure, but otherwise the ventral part of the postero-

internal columns are free from sclerosis, as are the postero-external columns. There is distinct but much slighter thickening of the neuroglia in the region of the transverse area of the cord occupied by the crossed pyramidal tracts, and, like the posterior column sclerosis, this change is limited to the cervical region of the cord. Apart from the affection of the posterior and lateral columns, no other areas of sclerosis exist in the cervical region, and the thoracic and lumbar regions appear to be quite free from sclerosis, even the parts affected in the cervical region escaping here. Throughout the whole length of the cord, as revealed by sections taken at different levels, a few scattered, recently degenerated fibres are seen chiefly in the posterior columns, and still fewer in the lateral columns. In the lumbar cord, however, there is a considerable increase in the number of these fibres in the posterior columns, where they occupy almost exclusively the postero-external tract. Beyond pigmentation, and staining black in sections treated by the Marchi method, the cells throughout the spinal cord appear to be intact.

Case II.—E. G., male, aged $15\frac{3}{12}$ years, was admitted to the National Hospital, Queen Square, on October 29, 1897, under the care of Dr. Beevor, complaining of trembling of the limbs.

Family History.—Patient's father, a mulatto from the West Indies, is alive and very alcoholic. His mother, a white woman, died at the age of 50 from phthisis. No relation, either on the father's or the mother's side, is known to have suffered from any nervous or mental disease. Patient is one of a family of six. The first child was born dead at full term, but "looked as if it had been dead for some time before birth." The second pregnancy resulted in twins, both born dead, prematurely, at the seventh month. Then a girl was born alive, but she soon developed "snuffles," and at the age of four years developed right hemiplegia and fits, both of which still persist. She is now 17 years of age, has a depressed nose, and there is a distinct history of her eyesight having been affected for some time when aged 8 years. Patient was the next child. He was born apparently healthy, and never had any symptoms of congenital syphilis. One other child has since been born, but it died of whooping-cough at 9 months.

Previous Health.—He has always been a strong, healthy child, exceptionally intelligent. At school he passed the seventh standard and carried off many prizes. He has never had any other illness whatever; no infantile convulsions. Three and a half years ago he was knocked down by a passing cart and

sustained a cut on his head. He was not unconscious at the time, and the wound promptly healed.

About three years ago his memory gradually began to fail. This was noticed first when he was sent out messages. He used to return having forgotten what he had been sent for. He also became much less bright at his lessons and ceased attending school. He was also noticed to "tremble" in his face and limbs whenever he was spoken to. Since then he has also had numerous spontaneous "trembling" attacks, during which the mental confusion has been more marked. There is no history of convulsions or twitchings of any sort. His memory and intelligence have deteriorated somewhat rapidly of late.

Condition on Admission.—Patient is a black-haired little boy. His complexion is very sallow. His eyes are prominent, giving him a somewhat startled expression, and his lips are large and thick. His hard palate is broad and normally arched. The teeth are well shaped and planted somewhat widely apart in front. There are no signs of congenital syphilis observable in the eyes or elsewhere. There is a small scar about half an inch long over the left mastoid process, about three-quarters of an inch from its tip, and there is a small horizontal depression on the mastoid process at this level, the result of the accident above noted. He can name objects readily, he can read and write, but in answering questions he is slow and stupid. His articulation is somewhat slurring and indistinct, and during talking there is marked fibrillary tremor in the muscles about the lips. He also licks his lips very frequently when talking, especially before beginning a sentence. He has no delusions or hallucinations; he is good tempered; he never speaks unless he is spoken to; he reads comic papers, but is very slow in appreciating the jokes. When, however, he does see a joke at last, he laughs loudly. He does not seem to care for play or amusement. His memory is very poor; he forgets which is his bed in the ward and tries to get into other patients' beds instead of his own. He has difficulty in repeating sentences read out to him, and tends to repeat only the concluding words of the last sentence read to him. When given a paragraph to copy out of a book he omits a number of words and mis-spells others. His hand-writing is tremulous.

He can smell asafœtida and peppermint normally in both nostrils, and he can taste sugar, salt, and quinine normally on both sides. A watch is heard at two inches distance in both ears. The fields of vision are normal on rough test; the optic discs and fundi are normal.

The pupils are dilated unequally, the right being much greater than the left. Both pupils are irregularly polygonal in outline. Neither of them contract either on exposure to strong light or during convergence. The external ocular muscles are normal; there is no squint and no nystagmus.

The facial movements, both upper and lower, are equal on both sides, but there is marked fibrillary tremor on voluntary movements, especially in the muscles about the lips and eyes. The fibrillary tremor is most marked when he opens his mouth widely. The muscles of mastication and the soft palate are normal; the voice is very tremulous. There is a coarse antero-posterior tremor of the tongue when it is protruded, not a true fibrillary tremor.

There is no anæsthesia anywhere to lightest touches or pricks on the face, trunk, or limbs.

All movements of the limbs can be executed well. There is no muscular weakness or atrophy. At rest there is no tremor, but on the slightest voluntary movement, as on holding his hand out in front of him, or on touching the nose with either index finger, there is considerable fine tremor of the whole limb.

The diaphragm and intercostals are normal, as also are the spinal and abdominal muscles. Patient's gait is somewhat ataxic and reeling; his back is slightly arched, and he stamps his heels in walking. He sways slightly on turning round rapidly, but can stand quite steadily with his eyes shut and feet together.

The knee-jerks are practically absent. It is difficult to be sure of their existence owing to the rigidity with which patient holds his legs in any position in which they happen to be. Occasionally, however, when the limbs happen to be relaxed, a very faint flicker of the quadriceps can be seen on tapping the patellar tendon, but not sufficient to cause any movement of the knee. There is no ankle clonus. The supinator-jerks, on the other hand, are extremely brisk on both sides. There is no jaw-jerk. The plantar, cremasteric and abdominal reflexes are brisk. There is incontinence both of urine and of fæces, but this appears to be due to patient's mental dulness, and not to any sphincter trouble. There is no onanism.

The external genital organs are very small, almost infantile. There is no pubic hair. The testicles are extremely small; only the left testicle is in the scrotum, the right is felt in the inguinal canal.

The heart, lungs, and abdominal viscera present no abnormality. The urine is normal.

On *December* 18th he developed scarlet fever, and had to be sent to a fever hospital.

Case III.—A. S., male, aged 17 $\frac{6}{12}$ years, was admitted to the National Hospital, Queen Square, on February 9, 1898, under the care of Dr. Ferrier.

Family History.—Both parents are alive and said to be healthy. There is no history of nervous or mental disease in any relation. Patient is the eleventh of fifteen children, of whom seven, including the two oldest, were born prematurely and dead, except one which survived a month. Of the eight full-time children, one died at birth, one died within a week after birth; three are alive and have no history of symptoms of congenital syphilis, two are alive but have had signs of congenital syphilis (one of these, a sister aged 14 years, had interstitial keratitis when seen on February 13, 1898). Patient is the only other full-time child. He was born apparently healthy ("a beautiful child") and was always very strong. He has had no illnesses whatever except measles. He is naturally of rather an irritable temper, but was clever at his lessons, passing the seventh standard when 13 years old. He never had any injury or accident of any sort.

About three years ago, when 14 years of age, he gradually became stupid and forgetful. If sent out on a simple message he would get confused and would return without having executed it. Then his actions became eccentric. On one occasion he took two caps from a draper's shop and brought them home with him; he also brought home an egg which he had appropriated from another shop. Ten months ago he bought a new knife and chopped up his watch with it, spoiling both knife and watch in the process. On another occasion he took off his trousers and in this state climbed on to the roof of the house. Within the last year he has become dirty in his habits, passing fæces in his clothes. Five months ago he was observed to walk unsteadily, reeling about but never actually falling down. About two months ago he seemed unable to speak for about an hour and a half—he "kept trying to talk, but could not utter any words." His articulation during the last two or three months has been noticed to be somewhat mumbling. He has never had any delusions or hallucinations at any time, and is said to sleep well.

Condition on Admission.—Patient is a well-nourished, fair-haired lad, height 5 ft. 2$\frac{1}{2}$ in. His expression is somewhat fatuous and his intelligence distinctly below par. When asked

how many pennies there are in a shilling he replies promptly, "Don't know and don't care!" On being asked a second time he replies "Twelve." He appears disinclined to answer simple questions or to perform any desired act. He refuses to attempt to write, exclaiming loudly, "*I* can't write, *you* write!" When asked to spell his own name he says "Artur." His voice is loud though rather tremulous, and his manner distinctly rude. His articulation is characteristic of general paralysis; it is slipshod, consonants being frequently elided. During speech marked fibrillary tremors are visible round the lips; his tongue is also very tremulous. The hard palate is well formed and not unduly arched.

He can smell asafœtida and peppermint in both nostrils. He tastes quinine promptly on both sides of his tongue, pronounces it bitter, and spits vigorously on the bed. Sugar he tastes, but calls it salt. His mental condition prevents accurate comparison of taste on the two sides. Hearing appears normal in both ears. The visual fields seem normal on rough test; the optic discs are normal.

The left eye tends habitually to be in a position of slight external strabismus, but he can voluntarily bring the eyes parallel, and can execute all external ocular movements well. There is no nystagmus. The pupils are unequal (the right measuring 4 mm., the left 3 mm.), not quite central. They do not contract during convergence, nor when strong light is focussed on them. The left palpebral fissure is slightly narrower than the right, but both upper eyelids can be voluntarily elevated to their full extent. The muscles of mastication appear normal. The facial movements, upper and lower, are executed equally on the two sides, but are accompanied by much fibrillary tremor on voluntary movement. The soft palate rises normally on phonation. The tongue is protruded straight, but exhibits coarse rapid antero-posterior tremor.

There is no anæsthesia anywhere to lightest touches or pricks on face, trunk, or limbs.

The grasp of both hands is rather languid, but whether this is due to want of will-power cannot be determined. He appears able to execute all movements of the upper extremities, but all movements are accompanied by a fine rhythmic tremor. There is no tremor of the limbs at rest. There is no rigidity and no muscular atrophy.

The abdominal muscles appear strong. He can raise himself in bed from the lying to the sitting posture without using his

hands. The spinal muscles are normal. Respiration is costo-abdominal in type, and normal.

All the movements of the lower limbs are executed normally as he lies in bed, but with the same fine rhythmic tremor as was observed in the upper limbs. His gait is feeble and unsteady, like that of an old man ; the feet are planted rather widely apart. There is no spasticity and no ataxy. He can stand with eyes shut and feet together. The knee-jerks are present and equal, rather subnormal. There is no ankle-clonus. The supinator jerks are brisk and equal. There is no jaw-jerk. The plantar and abdominal reflexes are brisk and equal; the cremasteric reflexes are absent. He occasionally passes urine and fæces in bed without calling attention to the fact, apparently from mental dulness.

The spine and cranium are normal. There is no tenderness on percussion, and no spinal curvature. The heart, lungs, and abdomen present no abnormalities. The genital organs are small for his age. Pubic hair is scanty. There is no axillary hair. The urine has a specific gravity of 1012; it is acid in reaction, contains no sugar, but has a distinct trace of albumin.

No signs of congenital syphilis can be detected in teeth, eyes, or elsewhere.

Patient's constant noisiness and shouting day and night necessitated his being discharged a week after his admission. Sulphonal, bromide of potassium and paraldehyde all in turn failed to quiet him.

Diagnosis.—The three cases described above were all undoubtedly examples of general paralysis of the insane. The differential diagnosis, when the occurrence of the disease in young people was borne in mind, presented no difficulty. A few of the possible alternatives may, however, be mentioned. None of the cases resembled cretinism in any degree save in the dementia, whilst they could not be classed amongst cases of congenital amentia, since all of the boys are known to have been possessed, previous to their illness, of intelligence above the average. Onanism, it is true, was observed in the first of the cases, but there is no evidence that it caused the dementia ; moreover, the diagnosis of gross central disease in this case was confirmed at the autopsy. The condition of infantilism, a rather

superior type of myxœdematous idiocy, recognised originally by Lasegne, and of which Brissaud* has recently described a case, differs markedly from the condition in our patients, inasmuch as all of them actually retrogressed after attaining a degree of mental development, and were not simply permanently arrested in their growth, mental and physical. In addition, they showed the clinical peculiarities of speech and articulation which are characteristic of general paralysis of the insane.

It may be useful here to give a short summary of the symptomatology of general paralysis of the insane occurring in childhood and adolescence, more especially as the condition is very inadequately described, even in the most recent text books of nervous and mental disease. The following conclusions are derived from a study of the records of such cases as I have been able to gain access to in contemporary literature. The best bibliography of the subject is in an excellent monograph by Alzheimer,[27] who gives a summary of nearly all of the cases recorded up to 1895, together with three of his own (forty-one cases in all). Since Alzheimer's paper, fresh cases have been recorded by other observers (*vide* Bibliographical List). In all I have been able to collect records of fifty cases, exclusive of my own three cases above described.

The age of onset varies from the ninth year upwards.

Out of 49 cases in which the age of onset has been recorded—

4	cases	commenced	between	9	and	10	years of age.
3	,,	,,	,,	10	,,	11	,,
4	,,	,,	,,	11	,,	12	,,
2	,,	,,	,,	12	,,	13	,,
9	,,	,,	,,	13	,,	14	,,
2	,,	,,	,,	14	,,	15	,,
13	,,	,,	,,	15	,,	16	,,
5	,,	,,	,,	17	,,	18	,,
1	,,	,,	,,	18	,,	19	,,
4	,,	,,	,,	19	,,	20	,,
2	,,	,,	,,	21	,,	22	,,

The commonest age of onset, however, is from the thirteenth to the sixteenth year, roughly speaking, about

* Brissaud, "Leçons sur les Maladies nerveuses," 1893.

the age of puberty. After the sixteenth year is past, the number of fresh cases becomes relatively much smaller as we approach adult life, amounting to only 25 per cent. of all cases occurring before the age of twenty-one. It would seem, therefore, that the age of puberty is a specially critical period in children of syphilitic taint.

Both sexes are affected, apparently with almost equal frequency (twenty-five males to twenty-eight females). Wiglesworth,[20] who recorded five cases, all of them in girls, was of the opinion that the female sex is specially liable to this form of brain disease. The comparison of a large number of cases shows, however, that his statement is only partially true, since both boys and girls appear equally liable. But this is in marked contrast to the overwhelming preponderance with which the male sex is attacked in later life.

The relation of the disease to syphilis has been well established, and in the great majority of cases a family history has been elicited pointing to inherited syphilis:—in thirty-six out of forty cases in which a family history was obtainable, making a proportion of 90 per cent. in which a hereditary taint was actually ascertained. These figures probably under-estimate the real frequency of syphilis in one or other of the parents of the patient. It is, however, noteworthy that objective evidences of congenital syphilis in the patient's teeth, eyes, bones and elsewhere are conspicuous by their rarity, the syphilitic poison in such patients seeming to expend itself chiefly on the central nervous system. Indeed, we sometimes find (as in Cases II. and III. of my own) that whilst the patient himself shows none of the ordinary stigmata of syphilis, his brothers and sisters may exhibit congenital syphilitic lesions of the most marked kind, although they escape the brain disease from which the patient suffers.

Acquired syphilis is a very exceptional antecedent, but it has been recorded in at least three cases. In two of these, recorded by Sommer[11] and Rey[6] respectively, the children were infected by a syphilitic wet-nurse, whilst in the third, recorded by Hüfler,[18] the patient, when a little

girl of five years, was infected by a kiss on the lip. (The symptoms of general paralysis in this case did not appear until the age of twenty.)

In a small number of cases there is a history of some fall or head-injury having preceded the onset of the disease (as in Case II. of my own), but it is doubtful whether such traumatisms have any influence beyond, perhaps, precipitating the onset of the malady in a patient already congenitally syphilitic. In other cases, again, a fall on the head has probably been merely an incident in one of the fits which sometimes usher in the disease, and frequently occur during its course.

The *Symptoms* of the disease may be classified into mental and physical.

Mental symptoms:—These typically consist in simple, progressive dementia in a child whose intelligence previously had been normal, or sometimes even above the average. The child becomes gradually dull, forgetful and apathetic, sometimes stuporose, but may have occasional bursts of passion. Grandiose ideas are, as a rule, conspicuous by their absence, therein contrasting markedly with the classical form of the disease in adults. Mild delusions and hallucinations may exist, but never of the extravagant type met with in adults.

Physical signs:—The patient's general development usually becomes arrested. Growth, which had previously been like that of a healthy child, either ceases or advances only very slowly. The genital organs remain infantile in type. In female patients, if the disease has set in before puberty, the catamenia remain absent throughout the remainder of the patient's life; if the onset of the disease has been after puberty, menstruation becomes irregular and soon ceases entirely. The general bodily condition, then, is one of arrested growth in an organism which had previously been normal in its development.

Fits or "congestive attacks" are common, and may be the first symptom of the disease. They may be simple attacks of unconsciousness without convulsions, they may be attacks of general "trembling" without loss of con-

sciousness, or attacks of speechlessness (as in Case III.), or they may be typical and severe epileptic fits (as in Case I.). Death frequently occurs in one of these congestive attacks, in uncomplicated cases.

Cranial Nerves.—The special senses—smell, taste and hearing—are usually unaffected. Optic atrophy with its accompanying impairment of vision is not uncommon. The pupils, as in the adult form of the disease, are often irregular in outline and unequal in size; the light reflex is generally sluggish or absent and the pupils may fail to contract during convergence of the eyes. Fibrillary tremors of the face, lips, and tongue are usually present when the disease is well established and are pathognomonic, whilst the articulation becomes typically slurring and slovenly, and the handwriting resembles exactly that of the adult general paralytic. In a few cases there has been facial irritability (Chvostek's symptom) on percussion over the seventh cranial nerve.

Sensory Functions.—The patient can usually feel light touches everywhere normally. General analgesia has been noted in a number of cases, *e.g.*, those of Hoch[34] and of Lührmann.[29]

Motor Functions.—Weakness of one or more limbs is very common and may be monoplegic, hemiplegic or diplegic in distribution. Such motor weakness often follows a congestive attack (see Case I.) and is increased by each successive fit. Unsteadiness of the upper extremities on voluntary exertion is common and may resemble the "intention tremor" of disseminated sclerosis. The lower extremities are frequently ataxic, sometimes, however, they are stiff and spastic, and in the later stages of the disease, when the patient is completely bedridden, permanent contractures may ensue (Thomson and Dawson[30] and others).

Reflexes.—The knee-jerks may be absent, or exaggerated, or normal, and this apparently whether the limbs be ataxic or not. (Thus in Cases I. and II. the gait was ataxic in both, yet in the former case the knee-joints were lively whilst in the latter they were practically absent.) The superficial reflexes are usually normal and the sphincters

are unaffected, although the latter may become uncontrolled through the progressive advance of mental dulness.

The duration of the disease has varied from six months (Lührmann[29]) to eight and-a half years (Turnbull[2]). Out of twenty-eight fatal cases ten lasted over five years and fifteen lasted over four years. Other cases were recorded as being still alive after four, five, and seven years of the disease. The average duration of the disease is, therefore, longer than in adults.

As the disease progresses the patient gradually becomes more and more enfeebled, the limbs tend to become fixed by contracture, incontinence of urine and fæces supervenes, bed-sores tend to form, and death may occur from exhaustion. More commonly, however, the fatal result is brought about by a congestive attack, and this even before the patient has become severely paralysed in the limbs.

Morbid Anatomy.—The morbid anatomy does not differ essentially from that of the adult variety of the disease. Changes have been found in the brain, spinal cord, and peripheral nerves.

In the brain chronic lepto-meningitis is the rule, with firm adhesions between the pia-arachnoid and the surface of the convolutions on the convexity of the cerebrum, usually most marked in the central and frontal regions of the cortex. The affected convolutions appear somewhat atrophied. Internal hæmorrhagic pachy-meningitis sometimes occurs, as in the adult form, with the occasional formation of a sub-dural false membrane. Dilatation of the ventricles is common, and frequently also granular ependymitis.

Microscopically there is well-marked sclerosis of the cortex, both peri-vascular and neuroglial. Fatty and pigmentary degeneration of the pyramidal nerve cells of the cortex of the affected convolutions is usually found. Similar changes, though less marked, may occur in the nervous and connective tissue elements of the cerebellar cortex and of the basal ganglia.

In the spinal cord various systemic degenerations have been described. These may occur in various columns of

the cord; for example, in the posterior columns, in the crossed and direct pyramidal tracts and in the ascending antero-lateral tracts.

Degeneration has also been found in the peripheral nerves.

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THE CEREBRAL CAPILLARY CIRCULATION.

BY JAMES CAPPIE, M.D.

THE thesis I venture to submit in the present paper is :—That metabolic commotion in the brain tissue is absolutely necessary to maintain an active capillary circulation through the cerebral vessels.

To support this heresy, as I suppose it will be called, I shall make use of a few of the most elementary principles in physics, and assume the soundness of one or two postulates. "Action and reaction are equal and opposite." "Pressure applied to a fluid is communicated equally in all directions." "Pressure by a moving fluid is in proportion to volume and velocity." "Movement takes place in the direction of least resistance." These will be my instruments of research.

I shall assume that the atmospheric pressure cannot directly bear on the contents of the cranial cavity, but that indirectly it can act on them through the blood-vessels that enter or make their exit through the skull. That pressure, therefore, must be directed, not, as in most other parts of the body, on the external surface of the intra-cranial vessels, but on their *internal* surface.

I also assume the soundness of the Monro doctrine as to the conditions that influence the mass of blood in the cranial cavity :—

"The ends of the lateral sinuses perforate the cranium at some distance from the arteries, that the returning blood might as little as possible suffer interruption in its course toward the heart. For any alternate stop of it, especially when occasioned by the stroke of the corresponding artery, must have had a worse effect on the brain than on other organs—not merely on account of the delicacy of the brain

and the thinness of the veins, but because being enclosed in a case of bone, the blood must be continually flowing out by the veins, that room may be given to the blood which is entering the arteries. For, as the substance of the brain, like that of the other solids of the body, is nearly incompressible, the quantity of blood within the head must be the same, or nearly the same, at all times, whether in health or disease, in life or after death, those cases only excepted in which water or other matter is effused or secreted from the blood vessels, for in these a quantity of blood equal in bulk to the effused matter will be pressed out of the cranium." [1]

According to Dr. Kellie, Monro was in the habit of illustrating his doctrine "by exhibiting a hollow ball filled with water, and desiring his pupils to remark that not a drop of fluid escaped when inverted with its aperture downwards."

Having simply formulated his doctrine, Monro did not deem it necessary to fortify his position by immolating a hecatomb of victims. He seemed to take for granted that anyone with a fair modicum of intelligence who would take the trouble to grasp his propositions would not be able to do otherwise than come to the same conclusions as himself. The facts were patent; the inference seemed inevitable.

By and bye, however, the experimentalist came, and with him confusion. According to Dr. (afterwards Sir George) Burrows, such "opinions" were "erroneous," and "these errors obstruct our advances in the pathology of a most important organ." He argued that "the principle of subsidence of fluids after death operates on the parts within the cranium as well as upon those situated within the thorax or abdomen," and that "experiments and physiological considerations lead us to the conclusion that the quantity of blood within the cranium is extremely variable at different times and under different circumstances." [2] In support of these "opinions" he made a series of experiments on rabbits.

[1] "Observations on the Structure and Functions of the Nervous System," p. 5.

[2] "On Disorders of the Cerebral Circulation."

A few months ago, if anyone wished to defend the position of Monro, it might have been necessary to enter on a lengthened argument. Fortunately, that does not now appear to be required. After having dominated and misled the opinions of physiologists for over half a century, an eclipse has come over the views of Burrows. Another experimentalist has come into the field, and after a series of elaborately conducted experiments, he has come to the conclusion that " in the living animal the Monro-Kellie doctrine is true."[1] A victory has thus been scored for common sense. At the same time it must rather be said that Hill's experiments have " completely overthrown " the views of Burrows, than that they have established the soundness of the Monro doctrine. The latter will ever stand securely on its own independent evidence.

In an essay like the present, I cannot enter fully into a historical detail of arguments for and against the opinion that the heart is not simply the dominant, but the sole efficient agent in maintaining the movement of blood through the vessels. There can be no doubt that at present the prevailing opinion is, " that the heart's action is continued onwards through the capillaries, and is sufficient to return the blood through the veins back as far as the heart."[2] It appears also to have become an article of physiological faith that in controlling local distribution, the vasomotor system of nerves is the all-efficient agent.

On the other hand, Professor Alison argued that " there must be some vital power concerned in *aiding*, not *opposing* the impulse from the heart . . . Local determinations are to be ascribed to a mutual vital attraction or affinity between the blood and the tissues of the body."[3] If anyone now ventures to defend the latter view, some expression of sarcasm will be the most likely comment. Nevertheless, not a little may be said in its favour.

On more than one occasion I have ventured to point out

[1] " The Physiology and Pathology of the Cerebral Circulation," by Leonard Hill, M.B., p. 35.
[2] " Cyclopædia of Anatomy and Physiology," vol. i., p. 671.
[3] " A Text-Book of Physiology," 5th edition, p. 219.

that the physics of the portal circulation may give important evidence in support of the Alisonian theory. My argument has been that as the blood from the portal vein has to traverse an extensive and minute plexus of capillary vessels before it can reach the hepatic vein, and so be sent on to the heart, considerable force will be required; that, in accordance with the law of pressure in fluids, that force must react as backward pressure on the blood in the mesenteric veins; that the walls of these vessels are not fitted to support such a strain; and that therefore, some supplementary agency that can act "in aiding, not opposing," the movement of the blood must exist in the liver itself. As opposed to this argument, I have not observed any answer that will bear a moment's consideration.

In the present essay, I have to submit further evidence in favour of the same doctrine from a consideration of certain points in the physics of the intracranial circulation. I have, however, a preliminary remark to make in regard to the mass of blood in the cerebral vessels. According to Professor Foster, the quantity must be "small, and making every allowance for rapidity of current, the interchange between the blood and the nervous elements must also be small." . . . In the rabbit, "while the weight of blood in the brain at any one time amounts to about 5 per cent. of the total weight of the organ, in the kidney it amounts to nearly 12 per cent., and in the liver to as much as nearly 30 per cent."[1] Now, this is surely a most misleading comparison. It is as if we were to take a mass of water flowing in a level marshy locality, and having wide banks, and comparing with it a fractional quantity of the same water flowing over a sloping bed and in confined banks, commend the former at the expense of the latter. Which of the two is endowed with the greater amount of kinetic energy? The one is of scarcely any mechanical value, the other can be used to drive powerful machinery. As between liver and brain the vital question is:—Which transmits the larger quantity of *arterial* blood through its vessels? No one will venture to

[1] "Text-Book of Physiology," p. 1134.

assert that the hepatic artery can convey to the liver more than a fraction of the amount conveyed to the brain by the two carotid and the two vertebral arteries.

For our present purpose we may take up the cerebral circulation after it has started from the circle of Willis. Without requiring to trace the course of individual vessels, we soon find the arterial blood flowing through the intricate meshes of the pia mater. On piercing the brain cortex the vessels are extremely minute, and their course is traced with difficulty. The blood, however, percolates the whole brain tissue, and we next find it flowing through the venous capillaries of the pia mater, and then through larger veins into the sinuses of the dura mater.

Several problems now present themselves for consideration. What forces maintain the flow of blood through the encephalic vessels? What are the conditions that influence the volume, velocity or direction of the blood currents, and thus determine the amount of intracranial pressure? How are the energising functions of the brain itself exercised? These problems are so intimately related that no one of them can be fully considered apart from the others, and it is almost a matter of indifference which we begin with.

In the first place, we may consider how the brain energises. If the sentiment "on earth there is nothing great but man; in man there is nothing great but mind" be true, then, in man's frame there is nothing great but brain. The activity of this organ is the condition of all conscious life. It affords the physical basis through which mental states and operations are revealed either to one's self or to the outward world. I have no intention, however, to make a survey of the whole circle of its potentialities. As bearing on the questions immediately before us, I shall restrict attention to one kind of activity —that associated with voluntary muscular movements; but if my argument here may stand the test of criticism, it will also apply to the centres of sensation and the field of association.

How, then, does a motor centre energise? The meta-

physician may here be envied the facile way he overcomes—or ignores—all difficulties. The Will wills a movement and movement results! But although ready to allow the reign of metaphysics to be supreme in mind as *being*, the physicist claims sovereignty in everything that refers to the brain as *doing*. He regards that organ as he would any other working instrument, and may investigate the conditions that may influence the storage, intensity and liberation of its energy. He does not require to puzzle himself as to the nature of the nexus between volition and the energising motor structures.

I shall assume that every group of muscles, when in action, is correlated to a definite energising centre in the brain, and for the sake of clearness I shall take a single simple action—that of lifting a weight.

When a weight is lifted by the hand a certain strain is thrown on the contracting muscles. The strain is equal on the tendon that acts as a pulley-rope and on the fibres attached to the bone that plays the part of a lever. So far, the mechanical conditions are well understood. But if we go deeper we find that the dominant moving power resides in a limited area of the brain mass. It becomes a point of interest to learn how a soft pulpy piece of matter can energise so powerfully.

When a current of electricity is being tapped from the accumulator, it is assumed the molecular vibration or commotion in the energising medium must be inconceivably rapid. No molar motion can be detected in the dead-looking heavy material of the medium, but work, compared to which that done by the strongest muscles is the merest trifle, can thus be accomplished with ease and precision.

It cannot be otherwise with the brain. Its energy must be molecular, and the degree of commotion among its molecules will condition the amount of work done.

From the remarkable ease with which the functioning of the brain is carried on, we are apt to form a very inadequate conception as to the extent to which purely physical forces are at work through its mass. In ordinary circumstances we are no more conscious of their immediate working than

of the weight of the quiescent atmosphere on the surface of the body. But when a blustering gale has to be faced we recognise the working of a potent energy; and when a gust of passion becomes a cyclone in the mental atmosphere, the existence of smouldering fires that may break out in volcanic explosion is revealed to us. In the exercise of muscular force we have special evidence of the powerful energy that may be dormant in brain cells, and we are driven to the conclusion that when a motor centre is in brisk action there must be intense commotion in its molecular structure.

But simple molecular activity can no more energise functionally than a string vibrating in a vacuum can produce sound. We must have metabolism as a whole in full play. This involves a sufficient supply of oxygenated blood, between which and the tissue there is an incessant interchange of material, and which requires every moment to be renewed.

The object of the circulation is here—as in every other part of the body—to assist in preserving the functional capacity of the organ by maintaining integrity of structure. It gives necessary stimulus in activity, and it affords material for recuperation after exertion. The supply of blood to individual organs has usually a direct relation to the requirement of the tissue, and unless proof to the contrary be afforded we may assume that the same principle holds good in the brain. Without deciding in the meantime how it is effected, I shall take for granted that the circulation will react sensitively to every change in the tissue. In sympathy with the degree of molecular commotion, it will be comparatively quiet or vigorously active.

If a man lifts a 56 lb. weight, and by sheer strength of muscle can whirl it round his head, and if the muscular force depends in the first instance on the energising of any part of the brain, evidently the strain within the latter must in some sense correspond to that in the muscles, in accordance with the law that action and reaction are equal and apposite. Molecular vibration must be at a high pitch, and the capillary circulation be correspondingly vigorous.

The movement of blood, of course, implies pressure, and from the extent of surface involved—from the myriads of

vessels that are taking a part—the sum of the pressures must be very great. First in the large arterial vessels; then in the arterial capillaries of the pia mater; next in the capillaries of the brain tissue; lastly in the venous capillaries and larger vessels, we have movements in a multitude of directions, and therefore a considerable number of antagonistic pressures. Then, remembering that all these movements and pressures occur in a cavity whose capacity is rigidly limited, we have in addition the pressure of reaction. When the fluids are strongly flushing the brain and the vessels on its surface, the resisting cranial wall will throw the pressure backwards into the cavity. Such combined action and reaction constitute *stress*. This will be equalised through the whole cranial cavity on account of the fluid contents being continuous. It cannot be greater at one point than at another. The pressures may be legion; the stress is one.

The degree of intracranial stress, then, will depend on the volume and velocity of the moving fluids within the cavity. But we are told that not only is "the quantity of blood in the cerebral vessels small," but that "the interchange between the blood and the nervous elements must also be small." If this be the case we have here a serious exception to one of the fundamental principles of physics. It would prove that the brain is really an indolent organ, or that we may have powerful energy liberated without any correspondingly powerful physical agency.

The analogy between the phenomena of combustion and those of metabolism may be carried a long way. Some comparison is made in perhaps every text-book of physiology, but it usually refers to the chemical rather than to the physical phenomena. An important analogy, however, may also be traced in their physical aspects. The engineer in regulating his furnace, or the cook in managing her kitchen range, knows that it is not a large mass of air surrounding the burning fuel that determines the degree of heat. It is rather a restricted amount of air which is allowed to percolate the fuel in regulated *currents*. These are strong, and they are independent of all propelling agency. The air is sucked into

and through the burning mass by an attraction greatly more powerful than the repelling force of the heat, and the rush may be strong enough to produce loud sonorous vibrations.

So with the cerebral circulation. Judging from the amount of blood that in a given time has to flow through the brain mass, we must infer that the rate of movement will be great. The quantity is to be determined, not by what can be squeezed out of the tissues after death, but by the number and size of the feeding vessels. Judged by this criterion it can be no "small" amount. According to the estimate of Haller, one fifth part of the whole mass of blood which leaves the heart is directed to the circle of Willis.

If any individual centre be small in bulk, its power will the more depend on intense molecular commotion, and if the capillaries are so minute that it is extremely difficult to trace their course, velocity of movement through them must make up for the limited volume of blood actually present.

The inference, then, seems inevitable, that when a motor centre is severely energising, its molecular structure must be in great agitation, and that the blood will be coursing through it with a strongly distending pressure.

But pressure, to be of any mechanical value, must usually be met by counter-pressure. We have seen how this is secured to the brain as a whole by the cranial wall, and the same principle will also apply to every individual centre. In the instance before us, where does counter-pressure or reaction come in? We have a moving power—the energising centre; we have a resistance to be overcome—the weight to be lifted; where is the fulcrum? If told it is to be found in the resistance afforded by the surrounding brain mass, we shall have to consider the conditions of stability there, but before discussing this point at present, let us look a little further.

When raising the pia mater from the brain surface, if a convolution be gently separated from its neighbour, a double plexus of vessels is seen to dip to the bottom of the sulcus. One set penetrates the cortex from without, the other makes its exit from within. On the surface of all the convolutions —in close contact with the dura mater—the same conditions

are found. It is evident that unless provision is made to prevent such an accident, a block will be very apt to take place in the circulation of the pia mater, and therefore in the circulation of the whole brain. I am not aware that any such risk has received the consideration of physiologists. And yet, if the pressure force acting on the circulation be purely mechanical—if it bears on the blood only from behind, through the circle of Willis—the risk will be very serious. When the brain is active—with its circulation at the fullest, and its pressures at highest tension—any impulse that might cause further expansion of its mass to an almost infinitesimal extent would inevitably lead to occlusion of the pial vessels.

Recurring to the motor centre, so energising as to cause a heavy weight to be lifted, we found an outwardly distending pressure occasioned by the high velocity of moving fluid to be present through the structure. We also found that to enable its energy to be liberated with economy and precision, the centre must do its work under severe repression. We now find that the distending pressure must be such that it will not be allowed to interfere with the circulation through the pia mater.

If now we take a more comprehensive view of the active brain, the importance of its circulation in a purely physical aspect again obtrudes itself. During the waking state, when energy is simply potential but special functioning is in abeyance, each and every part of the brain must be ready, at less than a moment's warning, to respond to its appropriate stimulus, and to take on itself its special duty. This implies a full and steady circulation. Let the neurons expand and contract as they may, let their protoplasm and the dendrons or filaments vibrate ever so intensely, no response of consciousness will ever be given without the oxydising and stimulating blood. To maintain functional power, therefore, every structure must, as it were, keep a certain *hold* on the circulation. It will thus be enabled to resist efforts to deprive it of its legitimate share, and it will also refuse to allow the tissue itself to be readily pushed aside. In this way every part of the brain may stand in the relation of fulcrum to any local centre that may be actively working. If the

hold on the circulation be relaxed, as will be the case during sleep, or after exhaustion, the stress through the whole cranial cavity will be lessened, and the working capacity of the brain itself be modified. Can all this happen if the blood circulates through the brain only under impulse from the heart? To say that it is extremely improbable seems to me to be a very mild way to express the negative.

The brain can be adequately studied only from a monistic point of view. Its functions and special centres are very numerous, but if the metaphysician insists that the unity of the mind must be regarded as an axiom, the brain in its successful working will be a veritable unit. Co-ordination, sympathy, harmony, must reign through every part. Between molecular activity and the capillary circulation, correlation will be absolute. They constitute one agency. The former may be the dominant factor, but if the exercise of function is to be full and exact, the response of the latter must be direct. As there is mutual interchange of material, so there is mutual attraction—mutual control—mutual dependence.

It is no more necessary for one to unravel all the subtleties of this mutual relation in order to prove its reality, than it is necessary for the physicist to explain all the secret transformations of energy that take place in his machine. Suffice it to say, that in the movement of sap in the leaf of a plant we have evidence that fluid and molar motion can be directly caused by molecular activity, and we have only to suppose that (for example) in the tissues of the brain the changes are immensely more rapid and powerful to recognise a legitimate analogy. When metabolism is in full play "the physical interchanges between the fluids within and outside the capillary walls, the chemical affinities, the attractions and the repulsions, the inconceivably rapid oscillations of the molecules, all go to constitute a force which draws the blood onwards and *actively* transmits it through the capillaries."[1]

Other reasons may be given why such a relation should

[1] *The Monist*, April, 1897.

be considered not simply likely but necessary. Obviously, the *ratio* of the heart's action must be the same to the whole interior of the cranial cavity. Its greater or less force or frequency cannot influence the determination of the currents through particular vessels. It may maintain a moving power through the larger arteries, but it cannot affect the distribution through the brain mass.

Again, and possibly this argument may by some be considered the most important of all, there appears to be an absence of vaso-motor control over the brain circulation. According to recent researches the tyranny of these nerves has been more than questioned. Their very existence in the most important region of the body is denied. "Baylis and I," says Mr. Leonard Hill, "have been entirely unable to find any evidence of a vaso-motor supply to the brain . . . . In a recent and exhaustive research, Gulland has failed to demonstrate by any known histological means the existence of any vaso-motor nerves in the pial vessels."[1] If such views shall be confirmed by other observers, they should afford a cogent argument in favour of Alisonian principles. If vaso-motor pilotage be dispensed with, what can possibly determine the direction of the circulating blood? Does it meander at its own sweet will, or is law still dominant, and must it move in the direction of least resistance? Does it simply *soak* into the tissues and then ooze out again? Do motor, sensory and other centres receive each a certain uniform quantity, apart altogether from the amount of work it performs, and, therefore, whether the need be great or small? To think so is to suppose that all physical law in the cranial cavity is suspended, and that metaphysic is as dominant in the brain itself as it is in the mind. The laws that affect moving fluids must here hold sway as absolutely as on the mountain top, or in the depths of the sea.

This leads us to point out an interesting and most important service afforded by the pressure of the atmosphere. One of the obvious purposes to which this agency is put is

[1] *Op. cit.*, p. 45.

to counteract the unfavourable influence of gravitation on the circulation of the brain when the body is erect. In the words of Dr. Kellie, it is "a force which must be constantly operating to maintain the plentitude of the vascular system within the head." As I have already mentioned, it bears on the internal surface of the vessels. It therefore acts on the arterial blood in the direction of the circulation, and it is opposed to that in the veins. So far as determination to and from the head is concerned, the pressure on one set of vessels is balanced by that on the other. Thus the pressure through the venous sinuses will not interrupt entry by the arteries. So long as there is no disturbing influence within the cavity, the balance will also be maintained through all the encephalic vessels. But if any influence at the capillaries may give rise to an *attractive force*, then, in relation to the atmospheric pressure, it will become *suction*, and unless physical obstacles interfere, the blood will be compelled to move toward the focus of attraction. The effect in this respect will be as certain as it would be on the fluids in the neighbourhood of an exhausted cupping-glass on the surface of the body. Toward any energising centre, therefore, the atmospheric pressure becomes an important adjunct to the action of the heart, and the direction of least resistance for the blood to move in will be *in the line of greatest attraction*.

The trend of physiology is so completely in the direction of *experimental* research, that an apology might almost be expected from me for presuming to submit what may be considered important issues without any attempt to verify my conclusions in the laboratory. "Such a subject as the hydrodynamics of the cranium," says Leonard Hill, "can only be investigated by experiment." I answer, as dogmatically, that by simply making use of those elementary physical principles that relate to moving fluids, and applying them to the peculiar conditions we find within the skull, more direct light will be thrown on cerebral physiology, and especially on the correlations of mind and brain, than will ever be made by the most careful experimenting.

Were scores of experiments made on animals after their death, which might seem to prove that by a force not greater

than that of the heart's action, some bland fluid may be made to flow from the carotid artery into the jugular vein, my faith in the Alisonian doctrine would not in the least be disturbed. It will be simply impossible to arrange conditions at all resembling those present in the living brain. The absence of strain, and of all the antagonistic pressures, would thoroughly vitiate any conclusions that might be drawn from such facts.

In regard to Mr. Hill's own experiments for the purpose of measuring intracranial pressures, they, too, are seriously vitiated by the simple fact that the animals operated on have previously been made unconscious. If unconsciousness exists, the molecular energies of the brain must be immensely reduced, and therefore the conditions that can influence active circulation will be in comparative abeyance. The principal, if not the sole source of intracranial pressure is the moving blood, and if the intimate relation of the capillary circulation to molecular activity be ignored, the results arrived at cannot be otherwise than partial and inconclusive. We get "the play of Hamlet, with the Prince of Denmark left out." The experiments have evidently been made with painstaking care, but from their very nature—from the impossibility of including the most essential factors—they will fail to give data more likely to throw light on the functioning brain than can be got without them.

FOUR CASES OF FRIEDREICH'S ATAXIA WITH A CRITICAL DIGEST OF RECENT LITERATURE ON THE SUBJECT.*

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SUMMARY:—*A group of three cases of Friedreich's ataxia; an isolated case; tabular comparison; remarks on some of the symptoms, ataxia of voluntary movement, static ataxia, Romberg's sign, nystagmus, choreiform jerks, athetoid movements, deformities of upper and lower extremities, and of spine, condition of sensory system, mental state, reflexes, order in which the symptoms appear; their mode of advance; period of onset; increasing complexity of diagnosis; diagnosis of isolated cases; diagnosis from chorea, from hereditary cerebellar ataxia; diagnosis as affected by indications of mental defect; diagnosis from disseminated sclerosis, from locomotor ataxia, especially in children, from cerebellar disease; pathological anatomy; theoretical views as to pathology of the disease; treatment; literature.*

THE list of family diseases of the nervous system is now fairly lengthy. Higier,[78] one of the most recent writers on the subject, makes out a list of thirteen groups, as containing the more important representatives, and some of these consist of several sub-groups. It is common to find, as Marie puts it, that each family even of fairly well-defined type such as Friedreich's disease has its own special traits; consequently the grouping of affected families is more or less arbitrary, and in many cases, as yet premature. As we shall see further on, the records of transition forms, connecting one class of disease with another, are multiplying. What is wanted is the continued careful recording of clinical cases of those rarer diseases with, if possible, exhaustive autopsies.

* A Thesis for the Degree of M.D. Edin.

Friedreich's ataxia is a well-marked clinical type; many cases belonging to it have a "family" character, many are "isolated"; only a few are, properly speaking, "hereditary." This paper first gives a clinical account of four cases of Friedreich's ataxia; then follows a discussion of a number of the more interesting points in connexion with the symptoms, diagnosis and pathology, along with a critical review of the recent literature.

A. A Group of three Cases of Friedreich's Ataxia in one Family.

The parents of the first three cases of Friedreich's ataxia about to be described were Irish. They lived in Ireland till after the birth of their fourth child. The father, a factory worker, was a heavy drinker, and died at 50 of liver disease. The mother, likewise alcoholic, died of pneumonia. She suffered also from gastric ulcer, obstinate pruritus, and eczema of face and neck. The eldest son informs me that his father used to say he knew of no nervous affection previously in their family. No history of insanity, fits or any other nervous disease can be obtained. The circumstances seem generally to have been very poor, such as are common to thousands of Irish in Dundee and Lochee. The patients belong to a family of nine, accounted for as follows:—

(1) First child, premature, died at two months.
(2) Hugh—*ataxic.*
(3) Mary—*ataxic.*
(4) Nora, married, died, aged 25, in childbirth; showed no ataxic symptoms; had three children, eldest alive and healthy, the other two died when a few days old.
(5) Maggie, married, subject to headache since her marriage; two premature children, dead; one alive and healthy.
(6) John, died aged 13 of typhoid; had shown no ataxic symptoms.
(7) Agnes, healthy, recently married.
(8) James, healthy (aged 17).
(9) Winifred, *ataxic.*

Numbers 5 and 8 show nystagmoid jerkings; their knee-jerks are normal. All were suckled by the mother. Mary shared her mother's milk with another child.

Case I.—Winifred M'A., aged 15, a mill-worker.

Patient's History.—She seems to have had good health until March, 1888, when she was sent into the Dundee Infirmary as suffering from typhoid fever. A brother, aged 13, had died at home of this disease. Her illness had begun with rigors and slight diarrhœa, and she had lain at home for three weeks before admission to the Infirmary. She was there found to be underfed, and to be suffering from slight diarrhœa, but beyond this there were no definite signs of illness, and she was soon discharged much improved.

A short time after this her father and eldest brother observed the first symptom of her present illness, namely, some difficulty in walking. Her mother also spoke of the way the girl kicked when asleep. Very gradually a change in her speech developed, so slight that it would probably have escaped notice, had not her brother Hugh been on the look-out for symptoms of the disease from which he and his sister Mary had long suffered. On the sensory side there were no complaints with the exception of some pains in the back of the thighs, occurring in the spring of 1894.

In July of that year I sent her into one of my wards in the Dundee Infirmary, and since then she has been more or less constantly under my observation. During the past two years the girl has frequently stumbled and fallen when running, and this seems to have been always in imperfect light, in the early morning or in the evening. Moreover, she says the staggering is worse when going to her work in the morning, both in summer and winter, rest having apparently a greater influence than fatigue in increasing it.

Present condition (February 21, 1897). Fairly well grown; expression of upper part of face bright and intelligent. The lower jaw usually droops and the mouth is partly open. Cheeks well coloured; nutrition rather below the average. (This is easily explained by her home conditions.) Bones seem well formed; muscular development rather feeble, but not specially so in any locality.

Nervous system.—The following conditions are to be noted on the *motor* side. When *sitting*, slight swaying movements of the upper part of the body occur; the hands can lie perfectly still. When *standing*, patient prefers to keep the feet some distance apart, and can then balance herself steadily enough if her eyes are open. She fixes them on some object in front of her. There is swaying of the trunk and head, made much worse

when the feet are brought close together. If the eyes are then shut, patient at once falls over to one or other side, or forwards. The tendons on the dorsum of the foot are seen to contract (*danse des tendons*), and the toes grasp the floor; the great toe, however, is often over-extended. Balancing on one leg is quite impossible. Her head is always bent forward a little, whether sitting, standing, or walking. Her *gait* is distinctly awkward and ungainly. The steps are of varying length, her feet are sometimes wide apart, sometimes they knock against each other; she cannot keep to a straight line. Her feet are brought to the floor with a slight stamp, through a jerky movement with the character, not of spasm, but of inco-ordination. The staggering character is more marked when walking backwards, and still better if the eyes are closed. She can turn round fairly well, and can stop quickly when told to do so.

All *voluntary movements* can be carried out in a way, but the *ataxia* which is so marked in walking is likewise manifest in other directions. For instance, when patient is told to pick up a small thread off a smooth wooden surface the hand and fingers oscillate a little, and she prefers to rest them on the surface before catching the object; there is an awkwardness about the act. When told not to touch the board, the hovering movement described by Charcot is shown. Apart from this ataxia, however, she can carry out her intentions—can button her clothes behind her back, thread a needle, etc. A full cup is slightly spilt when she carries it, but the oscillations are not increased when she puts it to her mouth. When put through other tests for inco-ordination in the upper extremities with the eyes shut, there is a decided defect. When told to touch the tip of her nose, for instance, her forefinger first reached the upper lip at its lower border. My notes show a distinct increase in the ataxia of the upper extremities since 1894. When lying down she can touch objects with her heel or toes without very marked inco-ordination.

Involuntary movements include some already mentioned—the swaying of the body, the oscillations in carrying out a voluntary movement. But there are also occasional choreiform twitchings in the face, especially drawing up the right side of the mouth, and some jerky movements of the same nature have been observed in the head and fingers. To this class, too, belong the evidently very marked jerkings that occur during sleep. Patient is quite unaware of them, but they must interfere considerably with the repose of her bed companions. They have been continuous since the onset of the illness.

Certain involuntary movements are seen in the eyes when they are strained by being directed to the extreme right or left, a slow nystagmus, about one oscillation per second. When the eyes are fixed on an object above or below the horizontal line, a very fine oscillatory movement can be detected.

When the tongue is protruded, constant slight movements are seen.

Disturbance of speech is to my ear decidedly more marked than it was in 1894. And yet it is not very bad, especially when patient is paying attention to it. Her voice is deep-toned, the words are uttered slowly, there is rarely any slurring; long words or combinations as "post office savings bank," "articulation," can be uttered with a single effort. A stranger might find little difference between her speech and that of other girls of her class, but my own knowledge and her friends' statements show some deterioration to have taken place. She can write legibly, but the writing is tremulous and uneducated; she holds the pen or pencil in a clumsy way, but one must remember that she has hardly written a line since leaving school three or four years ago. I do not think her writing has grown worse since I first knew her.

Sensory Functions.—Patient used to have a giddy feeling at times, but she is now free from that. There are no abnormal sensations whatever and the various forms of cutaneous sensibility are normal.

The muscular sense is affected; patient is a good deal astray in trying to localise the position of arm or leg when blindfolded. (In 1894 the sense of posture in the upper extremities was fairly correct.) She constantly makes mistakes in trying to distinguish between different weights with her legs, but with her arms is fairly correct. Hearing, sight, and taste are normal; deglutition, defæcation and micturition are normal. Patient has not yet menstruated.

The *knee jerks* are completely absent and have been since I saw her first. Superficial reflexes normal.

Condition of Muscles, and Deformities.—There is no sign of flabbiness and no muscular atrophy can be detected. But there is an increasing deformity about the hands. When the right hand is extended, there is a very slight flexion of the wrist, a slight hyper-extension at the metacarpo-phalangeal joints of the fingers, especially the fourth and fifth, and a gentle flexion of the inter-phalangeal joints. The little finger shows a decided bend forwards. The fingers can be well separated from one another, and the wrist moves freely in all directions when the fist is closed, but by no

effort can patient extend the phalanges completely. The thumb is not affected in any way. The same description applies to the left hand, though here the deformities are rather less marked.

This condition of the hands is very distinctly worse since 1894.

The feet show a very high instep and prominent dorsum when patient is resting, not so marked when standing. There is no talipes and no hyper-extension of the great toe beyond the occasional involuntary movement before mentioned.

A slight spinal curvature is present, convex to the left in the middle dorsal region, convex to the right in the upper dorsal and cervical regions. The bend forwards of the head is due to a slight kyphosis.

Grasp is good on both sides, and the muscular power of the legs seems to be normal. Patient is easily tired, but works for the full day, earning seven shillings a week as a shifter.

Electrical reactions are normal, both with faradic and galvanic currents.

Her psychical condition is normal; intelligence rather above the average.

Case II.—Hugh M'A., aged 33, a mill-worker, began to walk when 18 months old. Between his third and sixth year he suffered from some illness "like water in the head," and was for two years in bed. He went to school when 6, and remained there till he was 12, learning with normal facility.

At the age of 9 he had a bad fall on the back of his head, from a cart, and was rendered nearly unconscious, but there were no bad after-effects. On leaving school he began to work in the mill, and showed some nervous symptoms. Other workers were afraid he would fall among the machinery, and some suspected him of smoking. There do not seem to have been any ataxic symptoms sufficient to attract attention before this.

He soon got into the way of holding his head forwards, with his eyes bent on the ground that he might balance himself better; he preferred to keep his hands stretched out, the palms being directed forwards when walking.

From the age of 14 he found some difficulty in picking things up. There was not much change in his condition for the next four years, but he found that mill overseers were rather shy of engaging him in case of accidents. Between 17 and 19 he thinks he improved, but after that he became markedly worse, and from the age of 20 walking has been impossible. About that time he

had loss of sensation in his feet, and got a burn on one of his soles through standing on a hot coal. For a long time he has suffered from numbness and feelings of creeping and prickling in his feet and legs, not in the arms. Often about two hours after going to bed he has a sensation of blood rushing up to his head, and his heart seems to stop. His legs sometimes start involuntarily at night. He has had no lightning pains, no girdle pains, no vomiting, no disturbance of bowels, no affection of vision. From about the time at which he lost the power of walking he has had some difficulty in micturition.

On August 1, 1884, he was admitted to the Dundee Royal Infirmary, and though the notes then taken are brief and evidently imperfect, it is worth while reproducing them for comparison with his present state :—" Head hydrocephalic in shape, 22 inches in circumference. Dorsal spine shows right lateral scoliosis of very exaggerated character ; sternum raised ; left lower ribs touch crest of ilium.

" *Sensory Functions.*—Tendency to coldness in feet ; formication in soles ; tingling sensations in legs and feet ; anæsthesia to touch and pain below level of iliac crests ; pupils large, sight good, hearing good.

" *Motor Functions.*—Drop wrist well marked on right side when hand held out, muscles of fore-arm and ball of thumb flabby, diminution of grasping power, no power in lower limbs except slight power of extension of hips and knees.

" *Visceral Reflexes.*—Normal except that patient sometimes takes half-an-hour before he can micturate. No skin or tendon reflexes in lower limbs.

Slight tendency to waste in muscles of legs. Sweats a good deal."

In July, 1894, I examined him minutely, and on comparing the notes taken then with those made now, I find very little alteration.

State on Examination, February, 1897 (see *figs.* 1, 2 *and* 3).—An under-sized, fairly nourished man, face somewhat flushed, expression intelligent, features thick, this being especially seen in lips, head large, brachy-cephalic, 59·4 cm. in greatest horizontal circumference, held well forwards. His usual *attitude* is sitting in an arm-chair, with his arms resting on a table in front of him. If he has no table before him he likes to twist his arms round the arms of the chair, to keep himself from sliding off. When thus supported in a sitting position his left shoulder is depressed, and if the *spine* is looked at (see fig. 2) it is

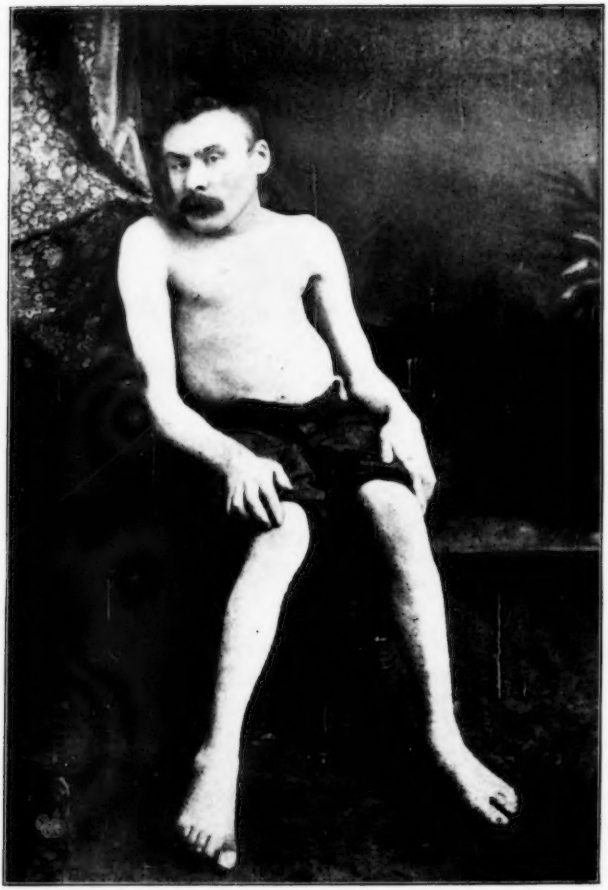

FIG. 1.

Case II.—General helplessness indicated. Patient is supported by man behind curtain.

seen to be curved (convex aspect) to the right, between the first and twelfth dorsal vertebræ. A slight compensatory curve the other way is seen in the lumbar region. The spine also shows a marked convexity backwards, in the upper dorsal regions, and a concavity (lordosis) in the dorso-lumbar. The upper four dorsal and the lower cervical spines are nearly horizontal.

When lying as straight as possible on his back the lower end of the sternum is very prominent, 7 cm. higher than the upper end. The sternum is straight, except the xiphoid, which is bent back. Intercostal spaces are narrow. No prominences at costo-cartilaginous unions. The hand can be easily passed behind the back on account of the lordosis. The ribs on the left are depressed to the iliac level.

Extremities.—(*a*) Upper. Muscles moderately well developed and pretty firm. No marked atrophy, though the interossei seem thinner and softer than any other set. No contractures, no fibrillary twitchings; no rigidity; passive movements possible in any direction.

(*b*) Lower. Muscles fairly firm, but considerably wasted, apparently from disuse. Right calf 25·6 cm., left 27 cm. in circumference; legs lie helplessly in bed, generally flexed at knee; foot shows a permanent deformity, the shape closely resembling that of *pes cavus*, arch extremely well marked on both sides, less on right than left; dorsi-flexion of first phalanges of toes, clawing of other phalanges to some extent. The deformity is less marked when the feet rest on the floor. On first exposing the right foot, a fibrillary tremor was noticed in the muscle on the inner border close to the arch (short flexor of great toe). Limbs very cold, and red or blue below the knees.

Motor Functions.—When in bed the patient prefers to lie on his right side, curled up. Cannot lie on left side longer than half an hour at a time. Has the greatest difficulty in raising himself, or turning round. In trying this he makes enormous efforts, painful to witness, and becomes very much flushed. The voluntary movements of the trunk are thus very jerky; he tosses himself round from one position to another.

Respiratory movements require a distinct effort; when sitting quietly with his table before him, both inspiration and expiration are audible through the room, a distinct pause separating them. The breathing is nasal; with each inspiration the head is jerked up a little. He is said to breathe more heavily and to snore at night. Power in the muscles of the neck seems well preserved; head can be moved in any direction. Occasionally there is a

slight choreiform vibration of the head, especially on making a voluntary movement. Swaying movements of the head and body are constant when sitting. No choreic twitchings of face, or other disturbance of facial movements.

Voluntary movements in the *arms* moderately active, but not so powerful as one would expect from the appearance of the muscles. Grasping power much diminished; free movement at shoulders, elbows and wrist. When told to straighten the fingers, the hand resembles the *main en griffe* of progressive muscular atrophy (see fig. 3). There is flexion at the wrist, over extension at the metacarpo-phalangeal joints, and flexion at the first inter-phalangeal joints. There is dorsi-flexion of the second phalanx of the thumb, and also of the ungual phalanx of the right mid-finger. (This the result of an accident.) The index is less affected than the other fingers, and the condition is more exaggerated on the right side. In extension the fingers are kept slightly apart, but they cannot voluntarily be completely adducted or abducted. The wrist and fingers can be quite easily straightened by an external force. The extensor tendons can be traced up the lower third of the fore-arm, and on the front the tendons of the palmaris longus and other flexors stand out. The palm is hollowed.

Movements of fingers and arms, in trying to pick up or touch objects, are badly co-ordinated. Patient likes to have the arms supported on the table, etc.; when this is done there is not much oscillation. Sense of posture in upper limbs defective.

As for the *lower limbs*, walking is quite impossible. Slight power of flexion and extension at hip. None at knee or ankle, very little at toes.

Sensory Functions.—In the legs there is slight diminution of acuteness in sensibility to touch, but pain is felt normally. No lightning pains; some slight paræsthesia in lower limbs is noted in the history.

Reflexes.—No knee or Achilles jerks, plantar reflex well marked on right, slight on left. Cremasteric and abdominal reflexes marked. Visceral reflexes mormal apart from delay in starting micturition.

Sight.—Normal; pupils act readily to light and accommodation; a moderate degree of nystagmus, consisting of irregular jerkings on fixing eyes on an object, to the side.

Hearing.—Slightly dull (this also the case in 1894). Hears watch at 12 in. on right side, at 3 in. on left. Occasional sound of rushing water in his head.

Tongue.—Flabby, marked by teeth, slightly furred on dorsum; when protruded a constant fine tremor pervades its whole extent.

FIG. 2.

Case II.—To show Scoliosis and Kyphosis.

FIG. 3.

Case II.—To show *main en griffe*.

Speech is markedly disturbed. One requires to attend closely to make out all he says, though if his attention is directed to words, he speaks much more clearly. The voice is monotonous, the words are somewhat slurred, not scanned. "Remember" becomes "rememr," "Whether" becomes "wher." "British constitution" becomes "Brish constution." "A dainty dish" becomes "a den dish."

Psychical state is normal. Patient is said to be rather irritable, and to make what the others consider undue demands from his being the eldest of the family. His intelligence is quite unaffected.

Electrical reactions with galvanic current. Extensors of left forearm KCC 7, ACC 12, KOC 8, AOC 9. Flexors of left forearm, KCC 9, ACC 9, KOC 12, AOC 13.

Heart—Sounds normal; position altered, apex being in third left space external to nipple.

Case III.—Mary M'A., aged 31, formerly millworker (figs. 4 and 5). *History:* Patient had inflammation of the bowels, measles and whooping cough in infancy, but there was nothing indicating any ataxia or paralysis until she was fifteen. At this age, in the month of July, she was taken ill one morning when on the way to work, feeling cold and suffering from bleeding at the nose and mouth. She became unconscious that night, and remained so for about six weeks. During the next eighteen months her mind was enfeebled, and she could not walk steadily; she managed, however, to do some work in the mill, and in course of time her mind recovered its former clearness. The staggering, however, increased to such an extent that at the age of twenty-one she was refused further employment. From the beginning of her illness she was subject to cramps and to jerkings in the legs at night.

At the age of nineteen, and again when she was twenty-one, she was a patient in the Dundee Royal Infirmary on account of spinal curvature.

Until she was twenty-six she was able to get about on her legs, pushing a chair in front of her, but during the last few years she has been unable to walk at all. She says that her hands have both been weak—the right more than the left—ever since the acute illness which marks the beginning of her disease. Still she could sew, although her hands were shaky; she has never been able to read or write, her school education having been very imperfect and short in duration. Since July, 1894, she had been almost constantly under my observation, in the Dundee Infirmary and the Parochial Hospital.

Present Condition.—A well-nourished, fresh-coloured woman, with cheerful expression, ruddy complexion and numerous freckles on face and neck. Her hair is light brown, strong and abundant; her nose is twisted from a fall four years ago, and on her right cheek is a scar, due to another accident. Through this scar there is a puckering of the cheek below the inner part of the right eye, when the lids are firmly closed; there is also a slight drawing-up of the right angle of the mouth.

The head is well-shaped, and measures 55·4 cm. in the greatest horizontal circumference; it is kept bent forwards through the spinal curvature, but patient says she was accustomed to hold it so because she was ashamed of her staggering.

Her usual attitude is sitting up in bed, with the back well supported by pillows; the spine shows a marked dorsal curvature, with convexity to the left, extending from the sixth dorsal vertebra to the second lumbar, the deviation at its maximum amounts to 1½ inches; the ribs project backwards to a marked degree on the left side, and are well separated from one another. On the right they are crushed together, and the twelfth rib is on a level with the iliac crest. In the upper dorsal region there is a slight curve with convexity to the right.

Some kyphosis is present in the upper dorsal spine; this can be removed partially, and for a very short time, by an effort of the patient.

The dorsal curvature is somewhat less pronounced when she is raised up by the arm-pits.

She complains a little of soreness, on pressure over the spine in scapular region.

The mammæ are well developed, and there is no striking asymmetry when the chest is examined from the front.

Extremities.—Upper extremities show, when at rest, no deformity; the bones are well shaped, the muscles seem smaller than normal on the back of the fore-arm. The lower limbs, on the other hand, show a deformity about the foot, talipes equino-varus being present on both sides. The legs below the knees are more cylindrical than usual, from a want of development of the calf; the antero-lateral group of muscles is deficient in bulk; the heel is drawn up, some transverse wrinkles of the skin being present over the lower end of the tendo-Achillis. The hollow of the pedal arch is very much greater than usual, and this is so even when patient is made to stand. Most of the grooves on the sole run from within, outwards and forwards. The great toe, especially the right, is always over-extended. It can be brought voluntarily

FIG. 4.

Case III.—To show *main en griffe* and deformity of lower limbs.

into line, but not flexed. The other toes show slight over-extension at the metacarpo-phalangeal joints; at the inter-phalangeal joints there is flexion. The dorsum of the foot is prominent.

Motor Functions. — Patient shows distinct weakness in the muscles that raise the *trunk* into a sitting posture. She can raise herself, but awkwardly and slowly. When sitting, there are constant swaying movements of the head and body; there are, besides, frequent jerkings of the head, and also choreic twitchings of the face, especially in the right levator anguli oris. These are not by any means so marked as to cause any confusion with true chorea.

In the *upper limbs*, movements are free at the shoulder, elbow, and wrist joints (with hand closed). When told to extend the finger and hand there is a marked imperfection; the wrist is somewhat flexed forwards, and to the ulnar side, the fingers are over-extended at the metacarpo-phalangeal joints (especially the ring and the little fingers), and flexed at the first inter-phalangeal joints. The terminal phalanges are in line with the middle. The deformity is, in short, the *main en griffe*. The thumbs are unaffected; the fingers can be separated, though less freely than normal.

Ataxia is present in the upper limbs, but patient can button her clothes, or thread a needle, if the elbows are supported. Her movement is like her sister's (Case I.) in picking up small objects, and in carrying out any movement oscillatory jerkings are very noticeable. The ataxia is brought out by the usual tests of bringing the fore-fingers together, or touching the point of her nose, with eyes shut. Hesitation and waving movement are more marked when she is near the point aimed at. The super-added movements are less marked if the eyes are open.

Power of grasping is still fair, though awkward from the extensor wasting. At elbow and shoulder muscular power is well preserved.

Lower Limbs.—Patient can flex and extend at hips and knees, but with lessened power. With her legs extended she can raise the right foot two inches off the bed, the left twelve. Even when supported on each side she cannot walk; her feet are pushed too far forward, so that she is unable to get her balance. Her steps are very short, the right foot is crossed over the dorsum of the left, and thrown two or three inches beyond the place where it ultimately touches the floor. She puts down her feet with a slight stamp, first the toes and then the heels. There is only

very slight voluntary movement at the ankle, and that is mainly extension. The toes allow of somewhat further voluntary movement. The ankle can be passively moved to a much greater extent, but dorsi-flexion of the foot is much interfered with, through contracture of the tendo-Achillis. The heel can be placed on the opposite knee, but slowly.

Sensory Functions. — Occasional headaches, no giddiness. There is some slight delay in conduction of impressions from the lower extremities, but sensibility in its different modes is essentially normal. There is a small area on the front part of the dorsum of the left foot, which shows slight dulling of sensibility to touch, not to pain.

Reflexes.—No elbow or knee jerk, abdominal reflexes well marked, plantar exaggerated, no disturbance of visceral reflexes.

Eyes.—Reaction of pupils normal; when looking to either side some nystagmus-like jerkings occur—about seven in five seconds, irregular in time. They are also seen when she looks up or down; sight is normal.

Hearing.—The hearing distance varies, but is usually about three inches on each side (*watch test*).

The Tongue, when protruded, is constantly in a state of movement—not the irregular jerks of chorea, but a fine oscillatory movement.

Speech.—Her voice is low-pitched, not harsh, monotonous. She keeps her teeth rather close in speaking. Some words are slurred—"docr" for "doctor," "I don't emember" for "remember." The speech is consequently somewhat slipshod; it is very like her brother Hugh's.

Vasomotor System.—Feet tend to be cold; psychical condition normal; muscular sense normal.

Electrical reactions. Faradic current much better borne on legs than normally; somewhat better on arms. Muscles of arms contract well under its influence, but the interossei act sluggishly.

Galvanic current.—In flexors of right forearm KCC 6, ACC 7; in extensors of right forearm KCC 6, ACC 7, KOC 8, AOC 8; in antero-lateral muscles of right leg KCC 6, ACC 10. The interossei, especially those of the third and fourth spaces, respond feebly to galvanism.

B. An "Isolated" Case of Friedreich's Ataxia.

Case IV.—William M., aged 13, admitted to Dundee Royal Infirmary, August 30, 1894.

Family History, &c.—Patient is third of a family of four, the

FIG. 5.

Case III.—Scoliosis. Patient requires to be supported.

youngest three by the same mother. His father died some years ago of inflammation of the lungs, after eight days' illness. His mother, sisters, younger brother, and nephew are all healthy. A slight nystagmus can be detected on extreme lateral deviation of eyeballs in his brother and younger sister. There is no history of any nervous affection in any of the other members of the family, nor is there any evidence of syphilitic or tubercular taint. His home surroundings are good; he has not been overworked at school, and has shown average intelligence.

Present Illness.—About two years ago his mother noticed that he became easily fatigued, and did not care to join in the games of his companions. His gait was a little unsteady; if he ran or walked quickly he was apt to stumble or fall. This was no worse in the dark, but if his feet were bare there was decided aggravation. There was no definite onset, no pain, shivering or feverish attack. His condition did not change much for some months, and then his upper extremities were seen to be slightly affected. He could not dress himself so quickly as usual, his hands were a little unsteady, and he was apt to spill the contents of a saucer in drinking. His handwriting was rather shaky, there was a want of freedom in using his pen or pencil and a difficulty in forming some letters. Both sides of his body seemed equally affected. The only time he complained of pain was a year ago, when his back ached at times, but this feeling passed off after the back was painted with iodine. He had no headache, or shooting pains in his limbs. During the past few months his arms and his legs have been getting thinner. No change has been noticed in his speech. There has been a steady deterioration from the beginning of the illness, the difficulty in locomotion increasing especially in trying to go downstairs.

State on Admission.—A fairly well-developed boy, with florid complexion, sad or pensive expression; brow transversely wrinkled, eyebrows often involuntarily drawn up while under observation. Weight 5 stone.

Nervous System (the only system affected).—The boy is quite intelligent, and has a keen sense of his unfortunate condition.

Motor Functions.—Patient stands with his feet about twelve inches apart, but if his heels are brought together he sways from side to side, and would fall if unsupported. Head and upper part of body bent forward. Closure of eyes increases the unsteadiness; when sitting in bed he has difficulty in balancing himself. Various muscles are seen to be constantly working as he tries to steady himself when sitting or standing. When laid on his back

he can raise himself up fairly easily by means of the trunk muscles alone. When standing he can stoop forwards, and pick any article off the floor.

Gait.—He walks with head bent forwards, and eyes fixed on the ground, his legs widely straddling. He brings his feet down heavily with a slap, first the toes, then the heels, reeling from side to side as he goes, and swaying his arms irregularly. His eyes are a great help to him in walking. He walks backwards with great difficulty; it is impossible to get him to keep a straight line. His gait has something of a festinant character; his steps are irregular in length, force, and direction.

Co-ordination, as tested by making him bring the tips of his fingers together, or touch other parts of his body, or place the heel of one foot on the opposite knee, is fairly good.

Writing is shaky; he presses his hands heavily on the paper. Nutrition of muscles, good; no hypertrophy or atrophy; no true fibrillar contractions, though muscular twitchings are noticed here and there even when he is lying at rest. No deformities about feet, but slight lateral curvature of spine, dorsal convexity and lumbar concavity to left.

Speech is low-toned and somewhat slow, not jerky nor scanning nor monotonous. The muscles of his lips seem slightly weak in their action, but there is no paralysis of the facial muscles.

There is distinct nystagmus (better marked on some days than others), brought out when the eyes are fixed on an object away from central line of vision.

Reflexes.—Patellar reflexes gone; no ankle clonus; plantar reflexes present on both sides, rather better marked on left; epigastric and cremasteric reflexes present; visceral reflexes normal.

Sensory Functions.—Has no abnormal sensations such as cold, heat, numbness or prickling. Sensibility to touch seems a little dull in lower limbs, the light touch of the head of a pin being frequently unfelt. Sensibility to pain acute, can readily localise a pin prick, can easily distinguish heat and cold.

Sight good; pupils equal, regular, reacting freely to light, but somewhat slightly to accommodation.

December 13, 1894.—Tongue thick and flabby, constant tremors in it, especially on protrusion. Frequent involuntary jerkings occur in head and upper limbs. Complains of feeling weaker, and often lies in bed for a day. Has occasionally been troubled with cramp in muscles of calf. His walking and general power of balancing are much worse, and he often falls. In trying to touch

an object he is more uncertain in his aim, from the irregular jerkings in his arms during voluntary movement. There is a corresponding difficulty in carrying out the heel-on-knee manœuvre.

Fundi of eyes show nothing abnormal. (Dr. Macgillivray.)

February 19, 1897. — Since last note patient has had fair general health. He has grown much taller, but there is no improvement as regards any of the symptoms described above. His speech is rather worse, the lips still seem somewhat at fault. The voice is more monotonous, and is low in pitch, distinct enough when speaking to me, but his mother says that when he is reading aloud to her, as he is fond of doing, she often has difficulty in understanding him. There is a thickness, a want of crispness about his utterance.

The ataxia of the hands is still as marked, if not more so. He can put in buttons, and write his name—always with the same awkward procedure. The superadded movements in using his hands are much as before.

He can walk, but only with strong support. His soles feel the ground quite well: the feet are raised higher than normal and brought down with a stamp, paretic not spastic. Romberg's sign is present.

When in bed he lies on his back; he has often a painful cramp, which causes the left leg to draw up of itself. The pain is immediately above and below the knee. The limb has first to be rubbed over the knee and then pushed down. If it were simply pulled down, the pain would be severe.

In the morning he has some pain in the lumbar region, when bending to get on his clothes with his mother's help. This is probably due to some stiffness, developed through his position in sleep. His back is very weak, and shows still a slight scoliosis; this is not of a permanent character.

His favourite position is sitting on the floor, with his back against the wall.

A recent symptom is a difficulty in commencing the act of micturition; there is no dribbling afterwards.

A brief *résumé* of these cases will be useful, and will show the main diagnostic points in Friedreich's ataxia.*

* (I take this opportunity of acknowledging the valuable aid given me in taking Cases I. and IV., by Dr. C. Kerr, then house physician in Dundee Infirmary, and by Dr. A. Croll, resident physician in the Parochial Hospital, in recording Case III.

Remarks on some of the Important Symptoms.

Disturbances of the motor system are by far the most important in the symptomatology of Friedreich's ataxia. So much is implied in the name, and Friedreich himself pointed out that the ataxia appears in two forms—ataxia or voluntary movement, and static ataxia. Choreiform jerks, nystagmus, tremors of tongue, and possibly disturbances of speech, are further evidence of wide-reaching motor invasion, while in the later stages of the disease certain muscles are apt to become paretic or paralysed.

Ataxia of voluntary movement is well shown in the gait. It has been often described, and has a character which marks it off from that of tabes dorsalis. In two of my cases (Cases I. and IV.) alone could it be examined. The patient in walking keeps his legs apart, and has difficulty in keeping to a line. He reels a little, sometimes a good deal, like a drunken person. The feet are brought down with a slight slap, first the toes and then the heels. The steps are short and vary in their length. The arms sway to some extent to aid in balancing, and for the same reason the eyes are fixed on the ground, a little in front; the head is bent forwards. In tabes the movements of the limbs are more disorderly, there is more excess of range, more exaggeration; the foot is raised higher, and brought down with more noise. The patient can keep to a line, if not asked to put one foot directly in front of the other. The character of the gait in each case varies, however, according to the stage of the disease, and, in tabes, according to the muscles worst affected. The point about the gait of Friedreich's ataxics which is most important from a diagnostic point of view, is its cerebellar character. This occurs also in Marie's disease, in multiple sclerosis ("rarely," Leyden), and of course in cerebellar disease.

But, added to the cerebellar or tabeto-cerebellar character, are various movements of the limbs and trunk best studied when the patient stands. These movements come under Friedreich's designation of *static ataxia*.* They are perhaps

* In one of Senator's cases (34) examined when the disease had made only very slight progress, the locomotor ataxia was very slightly marked, while the static ataxia was pronounced.

Case I.	Case II.	Case III.	Case IV.
(1) Family character.	(1) Family character.	(1) Family character.	(1) No family character.
(2) Onset soon after a fever, possibly typhoid, when 7 years old.	(2) Severe cerebral affection in infancy. Onset of present illness at 12.	(2) Acute illness (? meningitis) at 15, immediately followed by symptoms.	(2) Onset, insidious at 11.
(3) First symptom, difficulty in walking.	(3) First symptom, unsteadiness in walking.	(3) First symptom doubtful.	(3) First symptom, affection of gait.
(4) Very slow advance of ataxia; cerebellar character, slightly marked; worse in the dark (Romberg's symptom).	(4) Advance of ataxia, with remission, between 17 and 19; again rapid advance, with paralysis of legs at 20.	(4) Ataxia of gait, gradually increasing. Paraplegia nearly complete at 26.	(4) Ataxia of legs (tabeto-cerebellar), going on to complete inability to walk alone. Static ataxia. Romberg's symptom.
(5) Slight change in speech, beginning somewhat early.	(5) Marked articulatory defect.	(5) Articulatory defect.	(5) Speech only slightly affected and that recently.
(6) Slowly increasing ataxia of upper extremities.	(6) Ataxia of hands at 13.	(6) Ataxia of hands since the age of 15.	(6) Ataxia of hands and fingers, noticed after some months' affection of legs.
(7) Swaying of body; choreiform jerks.	(7) Choreiform jerks and swaying.	(7) Choreiform jerks and swaying.	(7) Constant swaying of body. Choreiform jerks.
(8) Nystagmus.	(8) Nystagmus.	(8) Nystagmus.	(8) Nystagmus.
(9) Absence of knee-jerks.	(9) Absence of knee-jerks.	(9) Absence of knee-jerks.	(9) Absence of knee-jerks.
(10) Slight weakness of interossei in hands, slight scoliosis. No talipes, or hyperextension of great toe, but unusually high instep.	(10) *Main en griffe* on extension of fingers. Kypho-scoliosis, very marked. Talipes equino-varus.	(10) *Main en griffe*. Kypho-scoliosis. Talipes equino-varus.	(10) No deformity of hands or feet. No permanent scoliosis.
(11) No marked paresis unless in hands.	(11) Great loss of power in arms, trunk, and especially in legs.	(11) Paretic condition in back and legs.	(11) Weakness of muscles, especially in back and legs; no atrophies.
(12) Intelligence and sensory functions (general and special) normal. Perhaps some impairment of muscular sense.	(12) Intelligence normal. General and special sensory functions normal. Muscular sense affected.	(12) Intelligence normal. Sensory functions normal. Muscular sense normal.	(12) Intelligence normal. Sensory functions normal.
(13) Visceral functions normal. Pupil reflexes normal.	(13) Delay in commencing act of micturition since age of 20; visceral reflexes otherwise normal. Pupil reflexes normal.	(13) Visceral functions and menstruation normal. Pupil reflexes unaffected.	(13) Visceral reflexes normal, till quite recently, when a difficulty in beginning the act of micturition came on (as in case II.). Pupil reflexes unaffected.

most strikingly shown when one looks through the camera at the patient. The inverted image is seen to be in a constant state of unrest, from swaying movements which are in inverse proportion to the amount of support afforded to the body. When patient lies in bed they are absent; when sitting with the back well supported by pillows, the head and neck move; if sitting unsupported, the movements are more marked, as the trunk is now swaying; and when standing unsupported, they may be seen at their maximum. A rough examination, then, shows the patient to be standing with legs apart, head and upper part of body bent forwards, tendons about ankle and on dorsum of feet in a state of perpetual motion, toes often with an athetoid movement, grasping the floor.

A more careful examination may show the various muscles of thighs, hips, and back in a state of irregular contraction, in the endeavour to preserve equilibrium.

Ataxia of movement is shown in the lower limbs when patient is lying by such common proceedings as putting one heel on the other knee. This test is carried out by Cases I. and II. very well, much better than one would expect.

Ataxia of upper limbs is easily brought out by writing, threading a needle, putting in buttons, picking up a small object.

The super-added oscillatory movements are present in carrying out any such attempt, and the movements are clumsy and awkward. The patient when lying seems to have more control over the test movements of the feet than of the hands, possibly because those of the latter are more delicate. Senator[35] notes that in his first case, incoordination is less marked in the lower extremities when lying or walking, than in the upper.

Case I. illustrates the bad effect of rest, for she is always worse as regards the ataxia in the morning, quite apart from the degree of light.

Tresidder[38] notes the same to a much greater degree in his Case I. He says "When 23 years of age, the unsteadiness in walking was so pronounced that a London physician was consulted, who thought that the lateral curvature of the

spine was producing paralysis, and the patient was ordered to lie down continuously. She did so for thirteen months. After this she began to walk again, and was at first very weak, but in the course of some months regained her strength until she could walk nearly as well as before the thirteen months rest."

Besold's second case,[54] a hawker, believed that his long walks (20-25 kilometers) were of great service to him.

These facts are of interest and importance, in view of the excellent results in the mechanical and gymnastic treatment of tabes attained by Leyden, Frenkel and others.

Organic changes are not affected, but the function of the muscles is improved by this method of "compensation-therapeutics." (Leyden and Goldscheider, Erkr. d. Rückenm, page 589.)

The test of making the patient close the eyes when standing or walking (Romberg's sign), while of very great value in locomotor ataxia, is not so useful in Friedreich's disease. Increase of unsteadiness through this is often absent, as in three out of four of Friedreich's own cases, in Rosenbaum's[83] elder case, in Bramwell's[12] first case. In my own cases that can walk (I., IV.) there is no doubt about the existence of this sign. In Senator's first case it was better marked on the second examination than on the first, a year and a half intervening between the two.

Where the patient can no longer walk, as in Case III., the same test can be applied to the upper extremities; here there is increase in the superadded movements when the patient is using her arms, with eyes shut.

The so-called *nystagmus* of Friedreich's disease seems best classed with static ataxia. It is not a true rhythmical nystagmus, in the sense of the ophthalmologist, though this occurs in rare cases (noted in three of those analysed by Griffith). The symptom must be carefully looked for, and brought out by directing the patient's gaze to one or other side. When the muscles of the eye-balls have their tension thus increased, distinct jerkings are seen—slow they may be and irregular, as in Case III., when the rate was seven in five seconds. Sometimes the movements can only be elicited

by turning the patients three or four times round, as in Geigel's[28] two cases (brothers, ages 20 and 18). On turning the patients to the right the eyeballs both turned to the right, and the short jerking movements of nystagmus to the left followed, lasting a quarter of a minute.

In two of Mendel's[10] cases, the movements thus brought out were very marked, and lasted some minutes.

They are found in most cases of Friedreich's disease—(" not in all " Gowers), but they are likewise often found in persons who have weak health, and even in the perfectly healthy.

This nystagmus I found in two quite healthy members in each of the families to which my cases belong, but no pathognomonic stress is to be attached to this.

On Schultze's instigation, Ausschlag[53] examined 200 unselected cases, and of these only 25 showed no tremor, though frequently there was only a single movement too far. " Only a strikingly high degree and special frequency of movements can be regarded as pathological."

Even modified nystagmus (" nystagmoid jerkings) is very uncommon in locomotor ataxia, so this sign may in certain cases be a help in diagnosis.

The involuntary movements, like an irregular tremor shown in the protruded tongue, may, without doubt, be referred to the same category as those we have been considering, and some would similarly treat the speech disturbances.

Besides the swaying movements of static ataxia, there are frequent *involuntary jerkings, choreiform in character*, but not so constant or violent as in chorea.

Thus in Case I., occasional twitchings are seen in the face, especially drawing up the right angle of the mouth. Very much the same facial movement is seen in her sister.

In Case IV., the brow is often raised involuntarily. The transverse wrinkles so marked in his brow may be associated with the bent position of his head, and may have been produced by his raising his eyebrows along with his upper eyelids. A waggling or wobbling of the head is constantly to be seen in the two worst cases, and to a slighter degree in the others. Even when lying at rest

occasional muscular twitchings, not fibrillar, are seen in Case IV. He is subject to a cramp in the left leg at night.

The other three cases have all been subject to jerkings in the legs at night. In the case of the eldest (Case II)., they seem to be more of the nature of cramps now, and another member of the family has often to get up to pull his legs straight.

The choreiform jerkings are an almost invariable part of the clinical picture of Friedreich's ataxia, and are of great diagnostic importance. They have sometimes led to a false diagnosis of chorea, as will be noted later on (*v.* Diagnosis).

In one case (Zabludowski's[84]) are noted "choreiform movements, especially when going to sleep," and also at that time, twitching in face and ears, and flexion of thighs on abdomen.

Painful cramp like contractions when in bed have been noted by others (Bramwell,[13] *Atlas*, vol. i., p. 46; Besold [54] Case II.). Nocturnal restlessness is noted in Bramwell's [12] Case I. Some of these exaggerated movements at night are probably analogous to the jerks which most people experience on dropping off to sleep.

Athetoid movements occur, too, sometimes in a quite minor form, as in one of Griffith's cases where was no tremor but a slow ataxic movement, resembling athetosis, which appeared when the hands were lying passively in the lap.

Athetoid movements of the toes in walking are occasionally present (Ladame's case,[1] p. 477). In both of Wallace Anderson's [36] cases, athetosis was distinctly marked in fingers and toes. These cases were peculiar in other ways; the eldest was twelve years old, but looked six or seven, and his mental condition corresponded to this age. The knee-jerks were present, though faint in both, and so was rigidity of legs with spastic gait. The onset in both was after measles two years previously.

The most remarkable movements of this class were observed in three cases reported from France. The first

(Chauffard[33]) was a boy of eight; symptoms since age of three; no heredity, sisters unaffected. No nystagmus, gait neither cerebellar nor tabetic. One leg made to pass before the other with a jerky and exaggerated convergent movement. Among usual symptoms were the progress, absence of jerks, absence of ocular symptoms, comparative preservation of muscular force, slight equino-varus. Further there was marked lordosis (dorso-lumbar), with slight cervico-dorsal kypho-scoliosis. No choreiform instability in repose, but sudden involuntary flexions of lower extremities at hips, legs remaining extended. Intelligence good, no grimaces. "Hovering" inco-ordination in picking things up. Arms took up athetoid position in walking with support. Forearms in position of pronation, hands flexed, their backs being directed down, fingers extended or flexed in the palm. The movements were not the usual slow convulsive movements of double athetosis. They only appeared when other voluntary movements were made—a muscular synergy, an athetoid synkinesia.

Chauffard easily separates this case not only from athetosis, but from tabes, insular sclerosis, cerebral diplegias, and chorea. But though there are several points of contact with Friedreich's ataxia, it is difficult to get over the uncommon features. The other two cases, reported by Londe and Lagrange,[72] are more like the ordinary type: Two sisters, aged 15 and 16, both took ill at six or seven. At first they were regarded as chorea. Absence of jerks, hollow-foot, marked inco-ordination, no nystagmus, internal strabismus in both; gait staggering; involuntary facial movement; head continually moving in various directions; choreiform jerks in head and limbs; *danse des tendons* in standing; slight scoliosis, no sensory or sphincter troubles, no affection of pupils or fundi; mental dulness. Athetoid attitude exactly as in Chauffard's case.

It is curious that in no one of these three cases was nystagmus present; in the latter two the internal strabismus, the mental dulness, and the athetoid attitude confer a special family mark.

Deformities of extremities and spine.—Certain deformi-

ties affecting the feet and spine have been always regarded as characteristic of Friedreich's ataxia. To these must be added the affection of the hand shown in my first three cases. None of these conditions is essential; the talipes (equinus or equino-varus) was noted by Griffith in only 27 cases out of 143. In a number of instances besides these there was dorsal flexion of all or some of the toes. The latter condition in itself might, as Besold suggests, be attributable to "modernes Schuhwerk," unless uncommonly well-marked. In Rütimeyer's families, Blattner and Kern, early prominence of the extensor propr. hallucis was regarded as a sign of very bad augury—it was one of the first symptoms (Ladame). Baskett's case,[76] a girl of 13½, was treated at a hospital for double talipes varus at the age of 8½. Her symptoms began when she was six, but usually deformity of the foot is of late occurrence. It is well-marked in Cases II. and III., but not in the other two. In Case I., however, there are indications in the high instep and occasional dorsi-flexion of the great toe that the shape of her foot will become like her brother's and sister's. The general appearance in a marked case corresponds closely to *pes cavus*, "hollow claw foot." The claw position may or may not be assumed by the terminal phalanx of the great toe. In Case II. (photo) the right great toe is over-extended as regards both phalanges. This is so, too, in Case III. In Plate X. in *Med. Annual* for 1895, representing a case under the care of Mackay, of Devizes, the great toes, like the others, have the second phalanges flexed, and this seems to have been the way in Ladame's patient.

Dreschfeld[50] describes a case which showed no talipes, but hyper-extension of the great toes "resembling that seen in the early stages of peripheral neuritis."

Deformities of the hands have been rarely observed. Bramwell[23] gives a family of three showing *main en griffe*. Griffith (*Amer. Journ. of Med. Sciences*, 1888, p. 377) describes one of his own cases as having "grasps strong but claw-like," another as having claw-like hands; but he may be simply referring to the mode of prehension so common in Friedreich's ataxia. In two of Rook's[7] cases, the hand

was claw-like and one had wrist-drop. One of Taylor's[55] cases had a little difficulty in extending the fingers.

In Cases I., II., and III., the attempt to extend fingers in line with the forearm at once brings out a more or less pronounced *main en griffe* (*see* fig. 3). Let me recapitulate the description of the condition in this case. When the hand is lying passively, nothing wrong is noticed. When he is told to straighten his fingers, flexion is produced at the wrist, with some bending towards ulnar side. The near phalanges are over-extended in the four inner digits, more so in the fifth than the fourth, in the fourth than the third, and so on. The middle phalanges are flexed, and the terminals are nearly in line with these. In regard to these joints, too, the deformity is more marked in the inner than in the outer digits. The fingers are separated a little from one another. The thumb is simply bent back. The more strongly patient tries to straighten the fingers, the greater does the alteration in form become. The extensor com. dig. tendons can be seen standing out on the back of the hand, and can be traced up the lower third of the forearm. The palmaris longus tendon, too, stands out prominently. The fingers can be adducted and abducted, but to a much less degree than normal.

What is the mechanism of this condition? It has nothing to do with the proper flexors and extensors of the wrist, for when the hand is closed, movement may occur at the wrist in the normal four directions—flexion, extension, ab- and adduction. It is not a paresis of the extensor com. digitorum; its tendons are inserted mainly into the near ends of the near phalanges, and Duchenne hence says they might be called *extensors of the near phalanges.* In the above case, so far from there being any sign of their failure to act, the chief trouble would appear to be their over-action (over-extension of proximal phalanges).

The cause is, I think, a paresis of the interossei—not paralysis—though the condition seems to come nearer this the older the patient becomes (compare condition in the three first cases). The interossei flex the first phalanges, and extend the second and third; they also abduct and

adduct the fingers. When they are weakened, and when the common extensor is thrown into action, the fingers assume the position above described. To keep the first phalanx in line with the metacarpals, antagonistic action of its extensor and flexor is needed. "When the interossei fail, the unopposed extensor causes an exaggerated extension of the near phalanges, and the extension of the middle and far phalanges does not only not occur, but they are, on the contrary, flexed in direct proportion to the degree of extension of the near phalanges." (Duchenne, *Syd. Soc. Trans.*, p. 265.)

The flexion at the wrist may be thus explained: when the hand is extended, not only do the proper muscles of the wrist act together, but also the long extensors and flexors of the fingers. The combined effect of their synergic action is to place the metacarp in line with the forearm. If the interossei are weakened, and the near phalanges are over-extended, the balance is upset, the common extensor acts at a disadvantage as regards its extending action at the wrist, and there is the appearance of a slight drop-wrist. This can be imitated to some degree in a healthy limb.

The greater amount of distortion in the ring and ulnar fingers corresponds to the condition in injury of the ulnar nerve, and is accounted for in this case by the escape of the first two lumbricals, which are supplied by a branch of the median. That the whole terminal part of the ulnar is not affected is proved by the intact sensibility, among other points.

The simpler case here considered leads to the consideration of the "Friedreich foot," about which a good deal has been written. I believe that it is analogous to the condition just described, and that its formation can be referred to an affection of the interossei of the foot (including the short flexor and adductor of the great toe). There may also be a real weakening of the antero-lateral muscles of the leg; there is certainly some retraction of the calf muscles, and tightening of the tendon-Achillis.

Paretic condition of various muscles besides the interossei is present in advanced cases. This is shown in the help-

lessness of Cases II. and III., especially as regards the whole of the lower extremities. The muscles that act on the spine may also be involved. In early cases, decided loss of power is usually absent.

Curvature of the Spine is very common in Friedreich's ataxia. Soca, quoted by Hallion,[24] says it occurs in half the cases. The photos of Cases II. and III. show the usual marked conditions. In the latter, the main convex curve is to the left, and is greatest about the ninth dorsal vertebra; in the former it is to the other side; it is both longer and more marked, and it is worst at the sixth or seventh dorsal vertebra. Perhaps the convexity to the *left* in the young woman is associated causally with the greater weakness in her right arm from her acute illness. In the common lateral curvature of adolescents, the main dorsal convexity is usually an exaggeration of the natural curve to the right, as in the brother (Case II.). In both there is kyphosis in the cervico-dorsal region, in the man this being so marked that the vertebral spines are nearly horizontal when he is sitting up. Observe how close the numbers on the spines appear to be, the effect of photographic distortion (fig. 2).

Hallion[24] has made a careful study of the spinal curvatures in nervous diseases, especially syringo-myelia, infantile paralysis, Friedreich's disease and locomotor ataxia. In all, the gray substance of the cord is affected in a different degree: in Friedreich's disease a pathological change is found in Clarke's columns, where some localise vaso-motor centres. Have we here, too, he asks, nervous lesions capable of causing an alteration of the bones, and specially of the vertebræ? He quotes Soca, to the effect that muscular weakness alone is sufficient to account for the condition. The want of tone may involve ligaments and bones too, as in the common scoliosis, but of this there is no proof.

We do not require to postulate any marked paresis of the spinal muscles; adolescents, who are subjects of the ordinary lateral curvature, are fairly muscular. A want of balance about the groups of muscles is more likely to start the scoliosis.

REMARKS ON THE CONDITION OF THE SENSORY SYSTEM.

The *sensibility of the skin* is frequently quite normal as regards pain, touch and temperature, but on the other hand it is not uncommon to find a slight dulling. In Cases I. and IV. the condition is practically normal; in Case III. there is some delay in recognising touch in the lower extremities, but localisation is correct. At one time there was absence of sensibility to touch on the dorsum of the left foot and the middle toes.

Case II. shows slight diminution of acuteness in recognising touch in the legs, but pain is felt normally; in the upper extremities sensibility to touch is normal. The curious thing about his condition is, that at one time (1884 and earlier) there was decided anæsthesia to touch and pain below the level of the iliac crest, this being proved by the examination in the Infirmary (1884), and also by the fact that about the age of twenty (1883) he burnt his foot by standing on a hot coal without feeling any pain.

It is interesting to compare this analgesia with the condition in the three cases recorded by Klippel and Durante, quoted in Londe's Thesis.[61] These cases are amongst the most important selected by Marie, from which to build up his type *hérédo-ataxie cérébelleuse*, but they are distinguished from the rest of this group, as well as from ordinary Friedreich's ataxics, by the disturbances of sensibility.

In Louis H., in 1892, tactile sensibility was abolished in legs and feet, much diminished in forearms and hands, and diminished in face; no plantar reflex; sensibility to heat recognised everywhere; retraction of field of vision. In 1894, Londe found analgesia in hands and legs; sensibility to touch present; sensibility to heat and cold weakened in the analgesic parts, retraction of field of vision hardly perceptible.

In François, sensibility to touch and cold, abolished below the knee in 1892, two years later no affections of sensibility.

In their sister, complete anæsthesia to all forms of sensibility on the internal aspect of the leg, in 1888; whereas a few years later, sensibility in her too, was normal.

Londe gives various reasons for his opinion that these sensory disturbances were due to hysteria. Certainly one is justified in regarding the condition as functional, and it has to be remembered, that many of the symptoms in nervous diseases, which have a well-marked organic foundation, are in all probability purely functional.

One of Stintzing's cases (*Münch. Med. Woch.*, No. 21, 1887) had complete anæsthesia in the lower extremities, and Senator's [35] second case felt on the feet needle pricks and differences of temperature, only slightly or not at all.

Déjerine [4] describes a brother and sister who were affected with marked sensory troubles of the extremities, diminishing towards the trunk, and also corresponding muscular atrophies; these he regards as true cases of Friedreich's ataxia, with very marked exaggeration of certain symptoms, sometimes met with in this disease, though only to a very slight degree. He refers them to lesions of the peripheral nerves and sensory roots.

It is interesting to note that Risien Russell, by his recent experiments (*Trans. Phil. Soc.*), showed that anæsthesia and analgesia occur in the partially paralysed lower extremities, after removal of the cerebellum.

Subjective sensations are also occasionally found. In Case II. these used to be, formication in the soles of the feet, and tingling sensations in the legs. Case I. had for a short time pains in the back of the thighs.

Lightning pains occurred in one of Déjerine's cases, very much the same as in locomotor ataxia. Bramwell [13] reports a family group of three cases, all of whom suffered from lightning pains at one time or another, one from gastric crises and all from exaggerated sensibility. As he notes, one of Charcot's showed lightning pains.

Other cases have involuntary spasms, associated with itching or other sensations (Tresidder's).

Mental Condition.

A normal mental condition may still be looked upon as characteristic. Tresidder's [38] three cases showed remarkably high intelligence, one of them having taken an

honours degree at Cambridge. Ladame's [1] case had a very vigorous memory and intellect. Hector Mackenzie's [48] was bright and intelligent for his station in life. My own cases are not affected mentally in the least degree. The *expression* of face is often unintelligent (as in Case IV.), and likely to mislead the observer. Cases that have normal mental power may be easily excited to uncontrollable laughter (Brock,[41] Ladame, Bramwell).

Mental defect is noted in some cases. In one case, epileptic attacks and dementia (Hoffmann, of Heidelberg [71]); in another imbecility (Power, quoted by Gowers, *St. Bart.'s Hosp. Reports*, 1882, p. 305); in two sisters, marked mental defect (Londe and Lagrange [72]); in one of Taylor's [55] increased mental irritability, with pronounced feebleness of intellect; in the other, not related to the first, epileptic attacks; in Nolan's [63] three cases, congenital idiocy; in Vinaj's [89] mental degeneracy.

In some of these, however, the diagnosis is more than doubtful. In itself it would not be surprising to find mental defect added to the usual symptoms; it would represent a further stage of the neuropathic tendency.

An American professor (Halbert,[68] Hahnemann Hospital, Chicago) gives a report of two cases of inherited syphilis with spastic-paralytic symptoms, as cases of Friedreich's ataxia, and in regard to the elder—eleven years of age—makes the quaint observation that "his mental condition is not greatly impaired, except that it is infantile from the lack of development!"

More will be found on this subject of mental condition under the heading of diagnosis (page 105).

Reflexes.

The condition of the *superficial reflexes* in Friedreich's ataxia may be regarded as normal.

In Case II. plantar reflexes are well marked on the right, slightly on the left. The cremasteric and abdominal reflexes are well marked. In Case III. all are well marked, and the plantar exaggerated. In Cases I. and IV. the superficial reflexes are normal.

In Berdez's[79] case, superficial reflexes were very lively. In three of Besold's cases[54] they were normal, in the fourth, abdominal reflexes were absent.

Variations in degree of cutaneous reflexes are of course of constant occurrence in healthy persons, and the point under discussion is chiefly of importance as helping to distinguish Friedreich's ataxia from tabes. In the latter affection the plantar reflexes, especially, are often absent, this being due to the loss of cutaneous sensibility.

Much more important is the *condition of the deep reflexes*, of which the knee-jerk alone may be considered here, as it is easily tested.

In the four cases I have described the knee-jerks are completely absent, and this is the case in the great majority of examples of Friedreich's ataxia.

Crozer Griffith (*loc. cit.*), in his analysis of a hundred and forty-three cases, describes the abolition of the knee reflexes as very early and very constant. In ninety cases only was their condition mentioned, and curiously enough in as large a proportion as twenty-two of these they were present, in six being actually exaggerated. It is true that Griffith's statistics are compiled from an unsifted mass of cases, but one can certainly not go as far as Ladame, who holds that the abolition of patellar reflexes is an absolute rule in this disease, and that one would do right to mistrust the diagnosis of Friedreich's ataxia whenever the knee reflexes are not lost, although all the other symptoms would seem to confirm it (BRAIN, vol. xiii., 1890, p. 492).

When one comes across a typical family of Friedreich's ataxics, such as that described by Tresidder,[38] and finds the knee-jerks completely absent in one and present in the others, one is forced to the conclusion that this test is not infallible. Moreover, it seems probable that the disappearance of the knee-jerks is, sometimes at least, not one of the earliest symptoms.

In Senator's [34] [35] first case, these were found to be simply weakened one year after the onset of the illness. After the lapse of another period of eighteen months the right was quite gone, the left could be obtained only by

Jendrassik's method. A sister of this patient, who had been ill for twenty years, had no patellar reflexes.

Brock's [41] second case, a man aged 27, had knee-jerks, though they were absent in his ataxic brother and nephew.

Erb [9] and Mendel [10] both showed cases in 1890 with tendon reflexes, in Erb's exaggerated. In one of Mendel's the jerks were present in May, but absent in the following November.

Londe [9] would class Erb's cases with Marie's hereditary cerebellar ataxia simply on account of the exaggeration of the reflexes, though vision was normal and talipes present.

In the cases I have quoted, as in two of Bramwell's,[22] the presence of the jerks may not be sufficient to blur the picture of Friedreich's ataxia, but it is a different matter when one finds that, along with retention of knee-jerks, there are other important deviations from the type. Bouchaud,[60] for instance, records a case which differs from Friedreich's ataxia not only in retention of the jerks, but in the impaired mental condition, hallucinations, affection of vision, nature of gait, and a too-pronounced nystagmus. I might refer further to the cases reported by Wallace Anderson,[36] Vinaj,[89] one of Nolan's,[63] T. S. Wilson's [39] first and fourth, as examples that are open to the above criticism.

However, the pathological anatomy of the disease varies, as we shall see later on, to such a degree that it is easy enough to suppose the knee reflexes might be free from interference in an occasional case. Such a case might be comparatively free from sclerosis in the lumbar portion of the cord.

Of the *visceral disturbances*, that noted in Cases II. and IV., viz., slow inception of the act of micturition, is not so very infrequent (thirteen times in Griffith's analysis).

Order in which the Symptoms Appear.

This is not easy to fix, but usually the ataxia of the legs shows itself first. This seems to have been the way in all my cases; in Case I. the speech was very soon involved, but so slightly that it would have remained unobserved,

but for the brother's thorough knowledge of the symptoms that might be expected.

In James's,[40] and one of Brock's[41] cases, the spinal curvature seems to have been the first symptom noticed. Both cases were lads of 18. One of Besold's[51] showed the ataxia first in the arms. In one of Mendel's,[10] the ataxia of the arms was noticed in the thirteenth year (writing becoming worse and more illegible), while it was two years later before the gait was observed to be changed. Griffith, in his analysis, states that the lower extremities were first attacked in 114 out of 143 cases, the arms in 10, and arms and speech simultaneously with the legs in 8.

Mode of Advance of Symptoms.

Some writers speak of the steady progress of the symptoms of the disease, and contrast this with the remissions of insular sclerosis. The mode of advance of the disease varies, however, to a great extent in different cases, and one may often distinguish remissions, or at least periods in which the symptoms are quite stationary.

Case II. showed slowly advancing symptoms till he was seventeen; then for two years there seems to have been a remission. At nineteen there was rapid deterioration; scoliosis and paresis, with unfitness for work, developed. Then the disease passed into its present chronic and almost unchanging state.

In Case III. the periods are less distinct; for years there has been hardly any alteration, but at about the same age as in her brother's case the extreme difficulty in walking developed rather rapidly.

The other cases (I. and IV.), show a steady advance, much more rapid in the latter than in the former. The age of nineteen or twenty will probably be critical in the history of Case I.

Period of Onset.

The date of onset of Friedreich's ataxia is by no means easy to state. Friedreich himself, judging from his own cases, considered that it was to be regarded as a disease of

puberty. Most cases undoubtedly begin before this, the seventh and eighth years being most common (Gowers[88]), and it is rarely that one commences after sixteen. The onset is so gradual, unless precipitated by an acute illness, that it may be almost impossible to definitely mark it. One of the latest cases seems to be Auscher's,—the woman being twenty-five when symptoms first appeared, but as we shall see, the diagnosis of this case must be accepted with reserve.

Londe (*Thèse*, p. 154) refers to two cases lectured on by Charcot, in 1893—brothers, in whom the onset was at twenty-five and twenty-one respectively.

In my cases, the fourth took ill about the age of eleven, but the beginning was insidious.

Case I. was seven when she had the illness, which seems to have been very soon followed by slight leg symptoms.

Case II. had a prolonged illness, possibly hydrocephalus, between his third and sixth year; there was, however, a long interval of good health before the ataxia became noticeable at the age of twelve.

Case III., like her sister, took ill immediately after a severe illness, though in this patient it seems to have affected the brain. She was then fifteen years of age.

The general statement holds good that it is a disease of the developing period.

Diagnosis.

The diagnosis of Friedreich's ataxia seems to become more involved as time goes on, and cases accumulate. In typical family cases, such as the three first above recorded, there can be little difficulty if one is at all cognisant of the group of symptoms that are required to define the affection. But the symptoms are by no means few, and not infrequently some are wanting. Hence, on the one hand there are close relationships with some other nervous disorders, and on the other more or less atypical cases occur, as to whose exact position there is disagreement amongst the highest authorities. And even in regard to some cases

where an autopsy has been made, and where very decided lesions have been found, there are differences of opinion; the clinical symptoms may put a case into one category, and the pathological anatomy into another.

Senator[35] considers that Friedreich's disease is a clinical conception, which would exist without any anatomical substratum, just as the notion of diabetes mellitus is independent of anatomical ideas. The pathological anatomy of some forms of diffuse myelitis ("combined sclerosis") is very like that of some cases of Friedreich's disease, but the clinical appearances are so distinct that no one could confound them.

The following case, one would almost think, had been diagnosed as Friedreich's disease after the *post-mortem* examination. It forms the basis of an elaborate paper by Tedeschi,[85] entitled "Die Friedreich'sche Krankheit — kritische und pathologisch-anatomische Untersuchung."

The patient, an orphan boy, aged 17, was admitted into the asylum at Florence in May, 1893. Two cousins in an asylum with mania. First symptom five years before, through a fright; tremor of right hand gradually involving arm, then left arm, next the legs. The tremor was like chorea. No pain; no remarkable affection of sight or hearing; no deformity. In asylum, increase of choreic movements, head involved. Walking difficult through incoordination, like an advanced tabetic. Speech slow and distinct; irritable, quarrelsome; of limited intelligence; could hardly understand what was said. Died in February, 1894, of tuberculosis.

One would like information about the knee-jerks, nystagmus, scoliosis, ataxia of upper extremities, and more about the sight and sensory system. But even with the details given—the mental affection, the uncommon onset, the asymmetry of onset, the absence of cerebellar type in the gait—I would have no hesitation in saying one does not, from the description, get the clinical idea that is requisite for the diagnosis.

The autopsy showed, besides tuberculosis in various thoracic and abdominal organs, a degenerative process in

the posterior ganglia and posterior roots; marked sclerosis of the posterior columns and Lissauer's zones (reminding one in extent and arrangement of the lesions in tabes dorsalis); marked atrophy of Clarke's columns and a slight change in the direct cerebellar tract in the cervical region. The cord was strikingly small, and there was chronic lepto-meningitis spinalis, specially posteriorly. The histological examination is most carefully described, but does not help the student of Friedreich's ataxia, if the above criticism is justified.

This, like Case IV., is a so-called "isolated" case—that is, he is the only one of the family affected with the disease. We are thus deprived of one important aid in diagnosis, namely, the "family" character.

Isolated cases are found to be not uncommon, and a survey of a large number in the more recent literature makes it clear that there are no outstanding characters which distinguish them from the true family cases occurring in collaterals. Divergences from the complete clinical type occur here and there, just as they do in the family form; and just as each family affected with Friedreich's disease shows its own special peculiarity (Marie [26]), so one must make a certain allowance for individual difference in the isolated form. This much being granted, there is still room for the criticism that in some of the published cases the narrator has shown a somewhat excessive desire to fit his irregularly shaped peg in the round hole of the "clinical picture." Take for instance the personally observed case given by Dana [17] in his article on Friedreich's disease in Keating's "Cyclopædia of Children's Diseases."

A lad, 18 years old, got an injury to the back of his head five years before. Soon he began to feel dull, heavy, and drowsy, had pains about knees and ankles, severe pains in thighs, pains in lumbar region, constant and severe cephalalgia, formication in legs, back, and shoulders. Stomach symptoms, vomiting occasionally, staggering gait, ankles unsteady. Felt as if tongue were tied back, consequently speech thick. Face became and remained flushed; much

dizziness; persistent polyuria (180-190 oz. daily, s.g. 1005-1010, no sugar or albumen). Diplopia.

So much for the history.

Physical examination showed a vaso-motor paresis of face and hands; "drunken" gait; no choreic or tremulous movements, except that head oscillated to and fro when he stood; slight tendency to flat-foot; no scoliosis; olecranon and *patellar reflexes normal*. Sometimes headaches, often vertigo; trunk ataxia; motor ataxia in using arms and legs; some nystagmus, which disappeared. Intelligence good.

Whatever this case is, it is not a typical case of Friedreich's ataxia. The history gives unusually copious and varied abnormalities of sensation, and as the author himself says, the origin from a blow on the head, and the cerebellar ataxia, are suggestive of tumour. The other symptoms all seem to point in the same direction (vomiting, polyuria, retention of knee-jerks, giddiness, &c.).

A case is reported by T. S. Wilson[39] (Case III.), as interesting from showing the symptom of glycosuria. The patient died at the age of 10 of diabetic coma, had been ill for five years, showed ataxia of gait, and slowness and hesitation in speech; a certain amount of clumsiness of hands; absence of knee-jerks; equino-varus; lateral curvature. There was no choreiform unsteadiness; no tremor on movement: great emaciation (from the diabetes, no doubt).

The case is certainly very interesting, and though the details are meagre, I should be more inclined to class it with the peripheral neurites than with Friedreich's ataxia. The sensibility was normal, but this may occur in a case of diabetic neuritis.

Case IV. of the same physician seems really to be what it was sent into hospital as, viz., St. Vitus' Dance. The unsteadiness became, in a fortnight from onset, so marked that the least thing would knock him down. There were muscular weakness; lordosis; increased knee-jerks; spastic tendency. Much improvement in hospital. Whole duration of illness to time of examination, only eight months.

Putting aside such cases as extremely doubtful, one finds considerable differences in those that remain.

Ladame's,[1] M'Caw's,[31] Baskett's,[76] Mackenzie's,[48] and Berdez's,[79] to name only a few, may be looked on as quite typical.

My Case IV. shows the essential points, namely, onset in childhood; first symptom (ataxia) in legs; progressive course involving legs and arms; affection of speech; swaying and choreiform jerks; absence of knee-jerks; Romberg's sign; nystagmoid movements; and amongst characteristic negative signs, absence of sensory defect; clearness of intellect; normal pupils; normal ocular fundi.

On the other hand, there are some points of divergence. Though the disease is now far advanced, there is no club-foot or extension of great toe and no marked scoliosis. Cases reported by Dreschfeld,[50] D. Inglis,[20] Besold,[54] (Cases III. and IV.), Zabludowski,[84] and many others quite undoubted, show no abnormality in this respect.

The hesitancy in beginning the act of micturition is not very uncommon, and is present in Case II. The cramp, curiously enough, affects only the left leg. According to Griffith, spasmodic contraction of muscles occurred in 21 of his 143 grouped cases, and was usually in the lower extremities.

Chorea is one of the diseases for which Friedreich's ataxia has several times been mistaken—Ladame, Londe and Lagrange,[72] Bramwell,[12] first case, one of Ormerod's (*Med. Chir. Trans.*, vol. lxviii., p. 149), &c.

It is to be distinguished by the slowly progressing character, the absence of knee-jerks, and the character of the involuntary movements. In marked cases of either disease there can be no difficulty. The tongue, for instance, in a choreic is jerked up and down, in and out, while in an ataxic it shows an irregular tremor with no rapid jerks. The face is usually not affected in Friedreich's disease, though in Cases I. and II. slightly marked exceptions may be noted. Chorea does occur in families. "Though the proportion is not great, the family tendency in some cases is very marked." (Gowers.)

Huntingdon's chorea is hereditary, begins in adult life, is accompanied by mental symptoms, and is always fatal.

Hereditary Cerebellar Ataxia.—It is by no means certain, as yet, whether this is to be looked on as a definitely marked disease with independent pathology. So far it would appear that the symptoms may vary to an extraordinary extent, and the autopsies on which Marie[46] based his reasoning amount to only two amongst the sixteen cases that he reckoned up as belonging to the new type.

Londe wrote a lengthy thesis on the subject (*De l'hérédo-ataxie cérébelleuse*), and added three new cases.

Marie recognises in all the character of staggering and cerebellar inco-ordination, and connects this with the cerebellar atrophy found in the two autopsies. He is supported in this by the most recent observations on the function of the cerebellum.

Both Friedreich's disease and the new type are family diseases, the latter beginning more commonly in adult life, the former in childhood. But as we have seen, Friedreich's disease may begin at twenty-five, and on the other hand, Fraser's patient (*Glasgow Medical Journal*, 1880) showed signs in infancy. This is one of the two cases of cerebellar hereditary ataxia in which an autopsy was made.

The various other points, when gone into, show a close parallelism in the two types. The symptoms of hereditary cerebellar ataxia may be summarised thus from Londe :—

(1) Slow and gradual affection of legs, in standing or walking.

(2) Occasional pains in legs and loins.

(3) Usually in three years or so, uncertainty of hands and arms.

(4) About the same time speech and vision involved. (In Londe's own cases the eyes were not affected, if one excepts nystagmiform movements).

(5) Reflexes remain or are increased. This is of course very exceptional in Friedreich's disease.

(6) Sometimes mental enfeeblement.

(7) Death at advanced age, usually from intercurrent disease, frequently of the lung.

It is much easier to say that the cases collated by Marie and Londe are not cases of Friedreich's ataxia, than it is to

admit that they should be put together to form a homogeneous group. But the theory put forward by Marie is at least plausible—that the same kind of degenerative process may be at work, involving the cerebellum only or mainly, in his cases—the spinal cord in Friedreich's disease.*

Relation of Mental Defect to Friedreich's Ataxia.

In cerebellar hereditary ataxia *mental enfeeblement* is often present. This leads one to consider the question "What weight should be attached to this point in making a diagnosis of Friedreich's ataxia?"

The pure type of this affection involves no mental change unless occasional irritability and a tendency to uncontrollable laughter; even this is exceptional. In fact, if we exclude the nystagmus and the speech affection, in regard to the causation of which we are quite in the dark, we have no evidence of involvement of the cerebrum, one might almost say, of the encephalon.

When the mind is affected we may take it for granted that the higher cerebral centres are diseased. We have seen that this condition is now and again present in cases that in other respects correspond to Friedreich's ataxia, but an examination of some cases recently reported makes one disposed to think that they should be regarded, not as Friedreich's ataxics, but as transitional to other types, *e.g.*, cerebral diplegia.

Amongst these I would reckon Nolan's[63] three cases of Friedreich's ataxia with congenital idiocy. These were a sister, aged 22, and two brothers, aged 15 and 10. Their mental condition was one of low intelligence; volition rarely manifested; memory defective; no delusions; speaking only in answer to queries; anxious to please; good humoured; asexual. All had nystagmus, internal strabismus; imperfect laboured articulation, many sounds not being uttered; ataxia in the arms and legs, with Romberg's sign.

* Compare Dr. A. Meyer's paper in the last (Autumn) number of Brain, with a reading of which I have been favoured by the Editor.

The thyroid was enlarged in the two elder. "Universal partial analgesia," and "drop-ankle" were present in the same two. Knee-jerks absent in oldest and youngest, exaggerated and combined with ankle clonus in the other. No scoliosis or Friedreich's foot; a "suggestion" of *main en griffe* in two; no visceral disturbances, or ocular troubles other than those referable to the external muscles. The true jerky or choreic movements do not seem to have been very marked. The disease was observed soon after birth.

There are indications here of an extensive defect of development in the whole nervous system. The cases seem to be more closely allied to cerebral diplegia than to Friedreich's ataxia.

Nearly related to these are the cases of a brother and sister (Bouchaud[60]), who at the ages of six and seven began to show mental enfeeblement, inco-ordination of the four extremities, and tabeto-cerebellar gait. No paralysis, nystagmus, or kypho-scoliosis. Speech slow but well articulated. The dementia steadily increased. Sight became lost; speech unintelligible; chewing and swallowing slow; rigidity of muscles increased, though knee-jerks were in one case absent, in the other diminished. The autopsy showed double sclerosis of lateral pyramidal tracts, diminution of sheathed fibres in cortex, anterior and posterior columns healthy; ganglion cells of anterior and posterior horn diminished in number.

The same category may be said to include Vinaj's[89] family, about which he asks "Is this a hitherto unrecorded kind of case or a modification of Friedreich's ataxia?"

Father, mother, and four children alive, second dead. Signs of nervous predisposition.

(1) Eusebio, well till age of 5, then difficulty in speaking and walking; intelligence affected, ending in idiocy; steady degeneration. When 11, paralysis of legs; paresis of arms; physical degeneration; no control of bladder or bowel.

(2) Marietta showed same symptoms at 5; died of accident at 9; by that time had lost the power of speech.

(3) Iside, age 6; slight nystagmus; knee-jerks normal;

gait uncertain; staggering; speech hesitating; scanning; memory weak.

(4) Egidio, aged 5, large head; unequal pupils; nystagmus; hesitating speech, pronunciation like a child of three; normal jerks.

(5) Healthy child of fifteen months.

In these cases the evidence against a diagnosis of Friedreich's ataxia consists in retention of knee-jerks, absence of talipes and scoliosis, and, most important of all, early and marked loss of intelligence. The family tendency, age of onset, ataxia, affection of speech, nystagmus and preservation of sensibility bring it into relation with that disease. The author rejects as diagnoses:

(1) Little's disease (origin; absence of special gait; absence of spastic rigidity; psychic phenomena).

(2) Spastic tabes.

(3) Primary progressive myopathy.

(4) Chronic encephalitis: primary cerebral sclerosis of infancy (time of onset; normal mode of birth; absence of convulsions, of contractures, of paresis or paralysis; the mental phenomena).

The cases are quite analogous to a number of others described by various authors, many of which are brought together by Higier[78] in his study of the rarer forms of paralysis of a hereditary and family character. After detailing four cases of his own in one family, he reviews the cases of cerebral diplegia reported by Sachs, Freud, Strümpell, Erb, Tooth, &c. The age of onset and the character of the affection are pretty constant in all, but each family has its distinctive characters. If one takes as the chief symptoms, onset in early childhood, spastic or flaccid paraplegia, idiocy, nystagmus, affection of speech and sight, one sees that Vinaj's and Nolan's cases are perhaps more nearly related to this group than to Friedreich's ataxia.

Higier's own cases are specially interesting, in relation to the point now under discussion, from the fact that they showed disturbances of co-ordination; weakness and awkwardness in upper extremities; intention-tremor; great

difficulty in sewing, knitting, and writing; in lower extremities weakness and stiffness followed by spastic paresis in gait, in one spastic ataxia for a time. He draws attention to the "little-known fact" that inco-ordination does now and then occur with increased tendon reflexes, quoting reports by Sachs, Haushalter, König, Dreschfeld. I may abridge the descriptions of Haushalter's cases:—Parents near blood relations, father alcoholic. Four brothers and sisters, normal births. Two eldest affected, now 11 and 10 years old. Symptoms first while at breast; rapid advance; inco-ordination of four extremities; fixed position of arms, of hands and fingers; slight rigidity in extremities, more in tongue and face; choreiform jerks when at rest; increased knee-jerks; *pes equinus;* anomalies of speech; disturbances of swallowing; imbecility. Nystagmus only in eldest.

These cases differ from Friedreich's ataxia in the retention of knee-jerks, the imbecility, the rapid advance at an early age and some other minor points, but they are transition forms between this disease and cerebral diplegia.

Collins[73] reports a case quite allied to those transition forms:—A child, 11 years old, of tubercular heredity (whose brother died in his second year, defective mentally and nystagmic), began to walk at end of first year. From third year gait stiffer, often falling; high myopia in fourth year; did not get on at school; when seen at age of 11, choreic movements of lower extremities; speech indistinct; exaggerated action of mimetic muscles under emotion; old face; gait cerebellar-ataxic, legs kept apart; ataxia of arms, especially left; increased knee-jerks; occasional ankle clonus; explosive ataxic speech; imbecile; sensibility and sphincters intact; no muscular atrophy; progressive paresis; slight nystagmiform movements; slight club-foot.

Here there is clearly a defective developmental condition of the nervous system in both cord and brain.

It is interesting to note how often in these cases which present a combination of the symptoms of Friedreich's ataxia with idiocy, the knee-jerks are retained. This tends to confirm the very high position of Westphal's sign as a diagnostic mark.

To the above list I might add the two cases of Londe and Lagrange,[72] who showed imbecility with the exceptional athetoid attitude in walking, and Hoffmann's[71] case, where there were imbecility and epilepsy, a case he himself records as doubtful in regard to diagnosis.

One sees from this review that when the degenerative process has involved not only the spinal cord, but the higher cerebral functions, other symptoms besides simple imbecility or dementia are almost invariably added to the symptoms of Friedreich's ataxia. And in such cases there must always be a doubt as to how far the changes in the cord, with their accompanying symptoms, are secondary to the cerebral affection, a doubt which a *post-mortem* examination might not clear away. Mental defect seems a much more important sign than the retention of the knee-jerk in making one doubt a diagnosis of Friedreich's ataxia.

Disseminated Sclerosis.

We pass now to consider the differentiation of Friedreich's ataxia from disseminated sclerosis. This disease is said never to occur in families (Marie), but a difficulty may arise when one meets an isolated case of Friedreich's ataxia. A good number of cases have been recorded of its occurrence in early childhood, but the proof of this anatomically was first given by Eichhorst in a paper "über infantile und hereditäre multiple Sklerose" (*Arch. für Path. Anat. und Physiol.* &c., Bd. 146, Hft. 2, p. 173). A well-marked case of insular sclerosis can present no difficulties; the intention tremor is coarse and jerky, much more so than in Friedreich's disease; the nystagmus is more pronounced; the course of the disease is liable to fluctuations in much greater degree; the tendon reflexes are exaggerated; ocular paralyses and affections of the optic nerve are frequent.

But the group of symptoms due to insular sclerosis varies much. The gait is at times cerebellar in type; knee-jerks are occasionally absent; scoliosis may be present; the speech may be similar to that in Friedreich's disease.

Brissaud (quoted in Londe's thesis) says it may be abso-

lutely impossible to distinguish an isolated case of *hérédo-ataxie cérebélleuse* from a case of *sclérose en plaques*, and the following cases may show the difficulty in diagnosing it from Friedreich's disease. Bouchaud [66] describes this one as " Maladie de Friedreich " :

Boy, aged 18, always rather delicate, fairly sharp at school. First symptoms at 12, viz., awkwardness of hands; tremors of upper extremities; difficulty in walking. Apprenticed to saddler at 14, but made no progress. On examination asymmetry of face (left smaller) ; left arm and leg more awkward and weak than right; intelligence much below normal; speech difficult to understand; does not speak much ; no choreiform instability when at rest ; static ataxia when trying to stand ; inco-ordination ; intention tremors; gait not tabetic; staggering, requires support; " tendency " to talipes equinus ; exaggerated extension of great toe ; knee-jerks both retained, left exaggerated ; nystagmus marked; slight lateral curvature ; acuteness of vision much diminished; hallucinations; no evidence of loss of sensibility.

This case differs from Friedreich's disease in quite a number of particulars, especially the mental condition, retention of knee-jerks, affection of vision, gait, asymmetrical appearance of face, greater awkwardness on one side. On all these points a diagnosis of disseminated sclerosis would be much more adequate.

Ewart [11] gives a case of a female, aged 19, of "isolated" type. Symptoms began at 16. Menstruation first irregular, then ceased ; slight affection of speech, only temporary, a year ago ; knee-jerks exaggerated ; slight tremor of hands on purposive movements ; slight lateral nystagmus ; marked ataxia of lower limbs ; slight loss of sensation ; functions of brain intact.

He thought disseminated sclerosis could be excluded by the insufficient degree of tremors and nystagmus and the normal cerebral condition. Friedreich's disease is, however, a doubtful diagnosis, on account of the disappearance of speech affection and the exaggeration of the jerks. These points would bring it nearer to disseminated sclerosis.

Probably, in a case like this, continued observation of the course would settle the question of diagnosis.

Locomotor Ataxia.

The diagnosis from ordinary locomotor ataxia can present little difficulty. A comparative table of the symptoms is given in Ladame's paper. A difficulty comes up when cases of Friedreich's disease have to be distinguished from tabes commencing in childhood. In the latter there is usually a syphilitic heredity, but not in the former. In a summary of ten cases Hildebrandt of Berlin (27) found that six began before tenth year, four before fourteenth. In some cases ataxia of extremities takes the leading place, lancinating pains in two, girdle sensation in four. Loss of patellar reflexes in all cases, eyes three times normal, nystagmus twice present, reflex paralysis of pupil twice, disturbances of sensibility in only half, disturbances of bladder six times. Moreover, the speech often shows disturbances on account of slight cerebral affection. Fortunately the symptoms are so grouped usually that there is not much difficulty in reaching a decision.

Raymond[91] discusses this subject at some length in a recent paper. He first records two cases, father and son. The father was a well-marked tabetic, showing, in addition to pre-ataxic symptoms, disturbance of superficial and deep sensibility, loss of knee-jerk, urinary incontinence, ataxia of the four limbs, characteristic gait, Romberg's sign, and marked eye affections. Onset in 1883, a few years after his marriage. Of his family, two girls and one boy were healthy, but the youngest boy had a nervous disease. He had always been rather weak in mind. In October, 1895, at the age of 15 he noticed one evening that he could not run, though he could walk. There was loss of muscular power; he soon found difficulty in climbing a ladder; he let things fall, and lost sensation of touch in his hands. He had no pains, but a sensation of wearing tight boots, even when his feet were bare. Gradually he lost confidence in movements, his gait became uneven, and his speech was embarrassed. In

January, 1896, this was his state: Development good; slight upper dorsal scoliosis; convex to right; tabeto-cerebellar gait; Romberg's sign; much impaired sense of touch in hands; impaired sense of posture; loss of deep reflexes; fibrillar and jerky tremors of tongue; nystagmiform jerks; frequent foolish laughter; intellectual functions limited. At present (September, 1897) condition rather worse; sight gone; (discs "tabetic"); jerky movements of limbs and trunk more pronounced, especially on effort.

In discussing this case, Raymond points out certain resemblances to Friedreich's ataxia, viz., gait, scoliosis, speech affection, nystagmus, jerkings, loss of knee-jerks. On the other hand, it differs in the marked affections of sensibility of skin and joints, and the optic atrophy. The case resembles tabes in these respects, in the loss of jerks and in the motor inco-ordination, but differs in the absence of a pre-ataxic stage with lightning pains, Argyll-Robertson phenomenon, urinary troubles; also in the swaying character of the gait, the nystagmus, the scoliosis and the speech affection. It is, in short, a hybrid case, a transition form between tabes and Friedreich's ataxia. Going into the question of juvenile tabes, he points out its great rarity; he himself has not met with a case in five hundred of locomotor ataxia. Only ten cases can he find in literature, with a doubtful eleventh, but these are all very characteristic, not differing from the adult form unless in a greater tendency to early development of optic atrophy, this being unaccompanied by any delay in the advance of the malady. Most of the cases published as juvenile tabes are cases of Friedreich's ataxia, or closely allied thereto, and amongst those Raymond would place two recorded by Freyer, and quoted by Hildebrandt in the paper above referred to.

But tabes in the adult may take abnormal forms. Auscher's[30] "case of Friedreich's ataxia," with autopsy, presents several points of close correspondence to locomotor ataxia. The onset was not till the twenty-fifth year; there was mental degeneracy, and abolition of light reflex. The patient was a prostitute, and there were no other members of her family affected. She showed marked ataxia of limbs;

choreiform movements of head and trunk; double equino-varus; loss of knee-jerks; nystagmus and affection of speech. Sensibility was retained, though in a case of marked mental defect it must be difficult to test it completely. Some lightning pains were present. The autopsy may be summarised thus:—Meninges normal; cord much diminished in size, like that of a child seven years old; cerebrum and cerebellum much smaller than normal, but free from microscopic changes; *posterior horns* much atrophied in whole length, worst in dorsal region; atrophy of fibres of Clarke's columns, number of cells in them diminished; *posterior columns* sclerosed in whole length, worst in lower part, Goll's columns being more changed than Burdach's; only a few medullated fibres in Goll's columns in a network of fine neuroglia fibres; *Lissauer's zone* normal in lumbar and cervical portions, perhaps slightly degenerated in dorsal; posterior roots and antero-lateral columns normal; motor nerves normal; sensory nerves showed a great number of non-medullated fibres, or fibres with an extremely thin sheath, perhaps embryonal.

The sclerosis showed what Déjerine had a few months before described as a pure gliosis: great abundance of fine neuroglial fibres arranged in whorls, absence of granule cells, no thickening of vessel walls or of septa connected with pia mater.

From the clinical account, as well as from the autopsy, one would prefer a diagnosis of locomotor ataxia, with an ill-developed brain, the congenital mental defect being possibly aggravated by a progressive paralysis, as Schultze suggests.[58] Criticism of a case like this is most desirable, for from it the following conclusions have been drawn:—

(1) Friedreich's disease may start as late as 25.

(2) Cerebellar defect may account for the clinical signs of Friedreich's ataxia (Senator).

(3) Mental degeneracy, abolition of light reflex and other exceptional conditions may occur in Friedreich's disease.

(4) The motor nerves may be normal, and the sensory show certain changes in Friedreich's disease (Leyden and Goldscheider).

(5) A full confirmation of Déjerine's views on the peculiar character of the histology in Friedreich's disease.

In making such inferences from a single case, the doubtful nature of the diagnosis must be borne in mind.

Disease of Cerebellum may cause symptoms like those in Friedreich's disease: reeling gait with a zig-zag character, difficulty in standing, nystagmus, loss of knee-jerk (usually not constant); but there is no tabetic character in the gait; the choreiform movements are probably absent; "the arms are usually steady, rarely they present some jerky inco-ordination" (Gowers), and in the case of tumour, pressure symptoms are generally present. Some cases of simple atrophy or hypoplasia of cerebellum have been described, but the symptoms are variable. In König's [65] case there was congenital cerebral diparesis, but no affection of inco-ordination at all, although the hypoplasia was very well marked.

"Oppenheim has observed cases of *cerebellar sclerosis*, and has had one anatomically investigated by Arndt. He regards cerebellar sclerosis as associated with a clinical picture composed of vertigo, cerebellar gait, ataxiform disturbances of movement, weakness of bladder, difficulty of articulation and motor weakness of the extremities" (Leyden and Goldscheider, *loc. cit.*, p. 597).

Although there is undoubtedly a general resemblance here, to cases of Friedreich's ataxia, there would probably not be much difficulty in distinguishing actual cases.

Cardossi [19] publishes a case of "*a rudimentary form of Friedreich's disease*," which he admits is by no means typical.

Giuseppe P., aged 40, has a son aged 18, who has had tremor in the right hand from the age of 13. He himself began to have tremor in writing at 10. Right arm and body affected; never so bad as to prevent him from attending to his usual occupation; gait, tabeto-cerebellar; has had phthisis for last ten years, not much advanced; oscillation of body when standing, brief, rhythmical, not well marked; intention-tremor in upper limbs, as in disseminated sclerosis; slight tremors of tongue; speech slow

and hesitating; no nystagmus; pupils normal; sensory system normal; knee-jerks much diminished; no paresis or paralysis; no deformities of feet.

The symptoms are of too mild a type and too few for Friedreich's disease of thirty years' duration, and they can be resolved into tremor, but apparently not that of chronic chorea, nor of paralysis agitans. Possibly a functional disorder allied to one or other of those.

In this discussion on *diagnosis*, I have not attempted to set forth exhaustively the differences between Friedreich's disease and the various affections likely to give rise to difficulty. My desire has been rather, by giving groups of cases from recent medical literature, to bring out the close relationships that bind one type to another. From a text-book stand-point this would be most confusing, but one cannot be too often reminded of the somewhat arbitrary character of the boundaries. The more numerous the doubtful cases that are published the better, though unfortunately there is sometimes a tendency to magnify this symptom or minimise that, so as to get the case included in a favourite type.

Pathological Anatomy.

The following are the more important records of *post-mortem* examinations in Friedreich's disease :—

(1-5) Friedreich and Schultze. (6) Everett Smith, *Boston Med. and Surg. J.*, 1885, vol. cxiii. (7) Newton Pitt, *Guy's Hosp. Rep.*, vol. xliv. (8-9) Rütimeyer, *Virch. Arch.*, vol. cx. (10) Letulle and Vaquez.[3] (11) Blocq and Marinesco.[5a] (12) Déjerine and Letulle.[2 and 6] (13) Gaizetti.[49] (14) Mirto.[37] (15) Michell Clarke.[57] (16) Burr.[74] (17) Dana.[77]

Autopsies by Menzel, Brousse, Kahler and Pick, Erlicki and Rybalkin, Rossi and Auscher, are rejected from the list as dealing with cases that can hardly be reckoned examples of Friedreich's ataxia.

For adequate reasons see Ladame,[1] Tedeschi,[85] and Schultze.[58]

Most of the cases that have been fully described have shown extensive system lesions. They have been summarised by Ladame in 1889, and more recently Tedeschi has given an analysis of 15 up to 1896. The most common conditions are diminution in bulk of the cord, sclerosis of posterior columns (especially Goll's), and degeneration of Clarke's columns. Next in frequency is degeneration of the direct cerebellar tract, and the antero-lateral ascending tract of Gowers. The crossed pyramidal tract may be partially involved, but it is thought that the true motor fibres are unaffected unless in very advanced cases. In one of Friedreich's cases the direct pyramidal tracts were involved. Apparent involvement of motor tracts is explained by the presence in them to a greater or less degree of neurons with other than motor functions. The " system " arrangement of fibres is, in short, not an absolute one. Occasionally changes in the spinal ganglia have been observed. The posterior roots have sometimes been found atrophied. Changes in form of the central canal, and sometimes peri-ependymal sclerosis, have been seen (Déjerine and Letulle).

Ladame's opinion that the zone of Lissauer (fine fibres at outer side of posterior column stretching from apex of posterior horn to periphery) always remains intact can no longer be supported. He looked on this as a point of distinction from tabes. But in several cases (Letulle and Vaquez, Blocq and Marinesco, Guizzetti, Mirto) it has been found more or less extensively altered.

The degeneration of Clarke's column, especially of its ganglionic cells, seems associated with the sclerosis of the direct cerebellar tract. The view that these cells are trophic centres for the tract fibres is thus confirmed. In early tabes Clarke's columns are affected. There is an absence of the fine fibrous network, while the coarse fibres, the ganglion cells, and the fibres of the direct cerebellar tract arising from them are unchanged. In advanced tabes Clarke's columns are almost entirely degenerated, and then the cerebellar tracts are said to be sclerosed (*see* Leyden and Goldscheider,[87] p. 509).

There appears to be no constant anatomical character

which would allow one to say from examination of a section of the cord that one had to do with Friedreich's disease, and not with tabes, or a combined sclerosis. Déjerine and Letulle have endeavoured to find a distinctive character in the nature of the sclerosis. Those authors believe that the sclerosis in the posterior columns is purely neuroglial, a gliosis, consisting of fibrillæ intercrossed in different directions, and forming whorls like those described by Chaslin in the brains of epileptics. Further, the pial prolongations into these columns are unaltered, and not more evident than in a healthy cord. In tabes, on the other hand, there is thickening of the septa of the pia mater, and of the prolongations which start from them, thickening of the neuroglial tissue between the nerve tubes, very pronounced changes in the vessels, &c. A vascular sclerosis such as this they found in the *lateral columns* in their cases.

To meet the possible objection that they are making an extensive generalisation from two cases, they point out that similar changes have been seen by several authors, but that they have not interpreted them in their true sense. They quote to this effect from the histological descriptions of Friedreich himself, Newton Pitt, Rütimeyer, and Blocq and Marinesco. Auscher, in his doubtful case, showed a similar condition at the Société de Biologie, July 26, 1890; and Déjerine, after examining his specimens, pronounced them examples of pure neuroglial sclerosis.

Schultze, of Bonn, re-examined by recent methods the cord of a case observed and described by Friedreich and himself in 1881, and he failed to find any justification for the distinction in the nature of sclerosis thus defined by Déjerine and Letulle. The whorls of fibrils were not present. The vascular thickenings were found in the posterior columns, especially between the columns of Burdach and Goll. He sums up his re-examination thus: No proper chronic meningitis. The degenerations are essentially a degeneration of the posterior roots and their connections in the posterior columns and posterior horns, disease of Clarke's columns, of the cerebellar fibres, and the pyramidal tracts—this so far as concerns the spinal cord.

Michell Clarke[57] describes the autopsy on a patient aged 14, a typical case except that the knee-jerks were normal, and that during the last few weeks of life he showed symptoms of cerebral tumour (cerebral vomiting, intense headache, optic neuritis, coma, &c.). A round-celled sarcoma was found growing from the under surface of the right half of the cerebellum, pressing on the pons and medulla. Apart from this no change in cerebellum. In cord no naked eye change except diminution in size. "No complete degeneration in any section, but a partial change chiefly in posterior columns, next most marked in lateral columns with involvement especially of the margin of the cord, with thickening in the pia mater and the walls of the vessels, and probably with some excess of the neuroglia."

A brother and sister were affected with the same disease. Had it not been for this, in view of the presence of a tumour in the cerebellum, one would have been apt to ask could not this have accounted for the symptoms. Certainly the symptoms were very severe at the end, and the tumour was apparently growing rapidly then, but it might have taken on an active character after a long comparatively latent course. The family had a strongly neuropathic tendency, and it is of course quite possible that two members of such a family should have Friedreich's disease and a third a cerebellar tumour without Friedreich's disease. Clarke himself states that the changes in the cord were by no means so marked as he was led to expect from the symptoms.

Dana's[77] autopsy is briefly recorded: it is accompanied by some photographs of cord-sections. In addition to smallness of cord, thickening of pia, sclerosis of posterior columns, sclerosis of lateral columns (crossed pyramidal and direct cerebellar tracts) and marginal sclerosis, he observed vacuolisation in the gray and white matter all through.

There is no note of the condition of Clarke's columns or Lissauer's tracts, nor could the author say whether the sclerosis was really a gliosis.

Burr's[74] autopsy was on a case that was eighteen

years ill. He found (1) marked degeneration of Goll's columns, slighter of Burdach's, from top of cervical to lower lumbar cord; (2) degeneration of lateral and anterior pyramidal tracts; (3) degeneration of direct cerebellar tracts. The sclerosis of lateral columns was not systematic. (4) posterior roots much degenerated, anterior intact; (5) slight affection of posterior horns with degeneration of ganglion cells, while in anterior horns only a very few cells were atrophied; (6) degeneration of cells in Clarke's columns; (7) walls of vessels slightly thickened.

The following are the chief points in Guizzetti's [49] case, who died of heart disease at the age of 28, after the nervous affection had lasted eighteen years: small size of posterior roots, spinal ganglia and cord.

(1) Lumbar region—sclerosis of posterior columns except a portion immediately behind posterior commissure and inner border of posterior horn, and also Westphal's posterior limiting zone; numerous degenerated fibres in pyramidal tracts.

(2) Dorsal region—posterior columns (with exception of part as in lumbar region), pyramidal tracts (numerous fibres), Clarke's columns and Lissauer's band all involved.

(3) Cervical region—Goll's columns as seriously involved as lower down, Burdach's less so, more healthy fibres in pyramidal tracts. Longitudinal fibres degenerated even into the nuclei of Goll's and Burdach's columns in the bulb, the horizontal fibres remaining unaffected. Some altered fibres even in anterior pyramids of bulb, contrary to usual condition.

Posterior horns atrophic. Microscopic condition like that described by Déjerine and Letulle.

In peripheral nerves, diminution of coarse nerve fibres and presence of numerous fine fibres.

Mirto's [37] case (death after five years' duration of illness, at age of 19) had systemic degeneration of posterior columns (Goll's, Burdach's and Lissauer's) of lateral cerebellar tracts, of Gowers' tracts; slight degeneration in crossed pyramidal tracts disappearing in cervical region, advanced sclerosis of Clarke's columns; rarefaction of

reticulum nervosum in anterior and posterior gray columns with partial cell-atrophy, partial degenerative neuritis in motor nerves (sensory not examined)—spinal ganglia affected as regards both cells and reticulum.

The latter two abstracts are taken from Tedeschi's critical digest, and I have not seen the clinical reports of these cases. But, supposing them to be typical, nothing of much importance has been added to what was known in 1889 to Ladame, or indeed Friedreich himself. The posterior roots and ganglia have shown marked changes, and in this respect and the coincident or consecutive affection of the posterior columns the similarity to tabes is obvious. In two cases (Guizzetti's and Mirto's) the principal nerves showed embryonic characters. In Mirto's the sensory were not examined, but the motor nerves were affected. Auscher's doubtful case presented similar appearances in the sensory nerves.

Pathology.

Various theories have been suggested to establish some causal relationship between the anatomical basis and the clinical symptoms. The general assertion that in a case of typical Friedreich's ataxia the symptoms are mainly spinal is one which has been widely though not universally accepted. As for the nystagmus, the affection of articulation, the giddiness and certain choreiform movements, though their origin is in all likelihood intra-cranial they have as yet no morbid anatomy. The strongest critic of the spinal origin of most of the symptoms is Senator.[34 35] He has maintained the view that the disease is essentially *cerebellar*, that in its most outstanding features it depends on an arrest of development of the cerebellum or of single physiologically important parts of it, and that it is only in a somewhat advanced stage that other symptoms appear, referable to super-added degenerations, in cord and cerebrum. The cord changes may be concomitant or consecutive. In confirmation he quotes Marchi's conclusion from experiments: "Destruction of cerebellum entails degeneration of

lateral portions of antero-lateral columns, of anterior part of direct cerebellar tracts and of a certain number of fibres in the pyramidal bundles." He says that we know nothing of the symptoms associated with disease of Goll's columns alone, that a separate or simultaneous affection of Burdach's would give a clinical picture like that of tabes dorsalis, and would be more likely to do so as certain other peculiar changes in tabes (affection of Clarke's columns, of Lissauer's zone, atrophy of posterior roots and degeneration of peripheral nerves) are not wanting in hereditary ataxia. Moreover, by reference to the autopsies he finds that the condition of the cerebellum was not mentioned in eight out of the fifteen he analysed. In three cases of the other seven there was a recognised atrophy (Menzel's, Rossi's, Auscher's). In three cases nothing wrong was seen microscopically, and in one nothing either with naked eye or microscope. This is the weakest point about the theory—the want of anatomical evidence—and this objection has not been removed by the later *post-mortem* examinations, though Clarke's case given above is interesting in this relation. As regards the three cases he quotes, we have seen how unusual in some important respects Auscher's case was; as for Menzel's, it is generally regarded as not a case of Friedreich's disease, and Rossi's also seems to deviate considerably from the classical type.

The small size of the spinal cord in Friedreich's disease seems to have struck all observers, and Friedreich and Schultze first advanced the view that the columnar degenerations were secondary to an *arrest of development in cord and medulla*. Shrinking of the cord no doubt occurs subsequent to the degeneration as the connective tissue takes up less bulk than the nerve tubes, but it is questionable if this would account for all the difference; it would hardly explain why the cord is so much less in this disease than in other scleroses, even if the points emphasised by Schultze are allowed their full weight. This author speaks of the general atrophy of the cord in tabes in the dorsal and lumbar regions as due to long disuse of the lower extremities. So in Friedreich's ataxia one would, he says, still more look

for such atrophy, on account of the inactivity enforced in childhood and the longer duration of the disease than in the case of locomotor ataxia.

The theory of developmental error is supported by other facts—the presence of numerous fibres without sheaths in peripheral nerves (Guizzetti, Mirto), anomalous development of central canal of cord (Déjerine, who quotes similar observations by Ormerod, Pitt, Everett Smith); condition of syringo-myelia (Friedreich's fourth case, quoted by Schultze).

No satisfactory explanation has yet been given as to why the cord should be able to perform its functions for a series of years varying from two to twenty or more, and then show signs of inadequacy. The demands made on the spinal cord of a young lad are great enough to test its capabilities most thoroughly. In certain families, and certain isolated cases, parts of the central nervous system seem to be badly constructed. They may be looked upon as unfit to bear a long strain, and their decay and death gradually come about; not that the change is analogous to a senile change; at least the differences are so great as almost to hide any analogy that may be supposed to exist. If the process of disease or decay has once been started, it is likely to be aggravated by the struggle for existence among the cells that is taking place constantly at all ages, and so actively in youth. The neuroglia rapidly increases, and not only fills the space formed by shrinking of the nerve-tubes, but compresses and destroys healthy nerve tissues (Roux and Weigert, quoted by Edinger).

Edinger[9] a few years ago brought forward his *repair* theory, to explain the condition found in several nerve affections. In any exertion there is waste of nerve-cell and nerve-fibre, compensated in health by repair. If nutrition is defective through any cause, as a toxin (syphilis, diphtheria), or injury, or hereditary nervous weakness, repair will be insufficient and degeneration may be the result. Some of the cord tracts seem more easily acted on than others, viz., the posterior columns, pyramidal tracts, and, less frequently, the cerebellar tracts. A greater strain may be put on certain nerve-lines than on others. Thus, taking up

tabes, he would explain the involvement of the nerve apparatus connected with the erect posture, the anæsthesia of soles (pressure on soles), affection of muscle sense, of eyes, &c.

In Friedreich's ataxia he thinks the progress of the disease may be explained as the legs and arms are used more and more with the progress of years. There is some abnormal weakness, either congenital or precipitated by an acute disease. The posterior and lateral tracts which are most used thus become affected. Here, however, Edinger admits there are serious objections to the adequacy of his theory. Why are other parts of the nervous system exempted through life? Why do the pupils, the bladder and the brain not show more disturbance than they do? No answer is attempted, but the very elementary nature of our knowledge of the physiology of certain parts of the nervous system is emphasised.

Dr. C. Macalister (*British Medical Journal*, April 8, 1893) suggests that the tendency to degenerate in certain diseases, such as pseudo-hypertrophic paralysis, may be due to the diminution in the functional activity of some gland whose secretion exercises a controlling influence over the nerve elements. Stawell [67] would extend the theory to cover cases of Friedreich's ataxia.

It will be evident that the above theories, however suggestive, do not by any means suffice to explain the whole disease in any large and general way. This symptom or that may be elucidated through the aid of other affections, perhaps more thoroughly understood. Atrophy of muscles, for instance, as of the interossei, may be referred to degenerations of their trophic cells in the anterior horns. Atrophies in this disease are not so extensive or frequent as to lead one to expect any very striking evidence of wasting of ganglion cells, but such wasting has been discovered.

The *ataxia* seems to be different from that of tabes, both clinically and pathologically. The cerebellar character may be referred, if not to the cerebellum, at least to the tracts conveying impulses to the cerebellum. The direct cerebellar

and the antero-lateral ascending tracts are longitudinal commissures between successive spinal segments and some part of the brain (Foster, "Physiology," par. 580). Another suggestion is that the posterior columns are the main tracts for conveying motor impulses downwards (Inglis, *Journal of Nervous and Mental Diseases*, January, 1892). Hughlings Jackson's view that cerebellar ataxia is due to weakness of the back muscles, and to the consequent attempt of the legs to correct the weakness of the back, is strengthened by such cases as those we have been considering. (For valuable discussion of this subject see paper by Ormerod, *British Medical Journal*, 1894, vol. ii., p. 1291.)

But the ataxia is not only cerebellar, it is tabetic also. The muscle-sense to whose impairment the most widely accepted theory refers the ataxia of tabes, is usually, though as we have seen not invariably, well preserved in Friedreich's disease. With a pathology so alike, as regards the posterior columns, one is tempted to think that whatever explanation is adequate for the ataxia and absence of jerks in tabes is likely to be sufficient *pro tanto* in the case of Friedreich's disease.

Therapeusis.

Almost all writers on this part of the subject speak in the most hopeless tone. However, Zabludowski in his patient, a little girl of 9, obtained decided improvement by a four weeks' course of massage. The case was an isolated one, and was exceptional in some minor points; for instance, regurgitation of fluids through the nose occurred in swallowing, formerly solids also had been thus returned. The "cure" so improved her that she seldom fell, walked straight, and could feed herself without letting the spoon drop. Massage increases the functional capacity not only of healthy muscles but of those whose activity is impaired by direct or indirect causes. The nerves are better nourished through the improved circulation. Nerves and muscles only partially diseased may take up the work of those that are further gone. To this one might add Weigert's concep-

tion of the mode of action of a stimulus. "Neither cell nor organ is directly strengthened by the stimulus; the apparent strengthening is due to the fact that in action portions are used up and replaced by new fresh portions, a substitution which in many cases occurs in excess." (Edinger.[90]) Improvement thus obtained can hardly be anything but temporary. Case IV. had a month's massage in hospital but without benefit. It is clear, however, that rest is the worst thing possible for such cases; they should be encouraged to take exercise, and to go through such actions as will best bring their co-ordinating faculty into use. Touching definite points, picking up small objects and placing them on marked spots, writing, walking without or with aid—these and other exercises employed in the mechano-therapeusis of tabes should be useful in Friedreich's ataxia.

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(84) ZABLUDOWSKI. "Ein Fall von Friedreich'scher Krankheit. Behandlung durch Massage." *Berl. Klin. Woch.*, p. 757, 1896.

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(86) MAGNUS-LEVY. *Deut. Med. Woch.*, No. 28, 1896. *Ver. Beil.*, No. 19. Isolated case, doubtful. Details scanty.

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A Case of Friedreich's Ataxia, with Autopsy and Histological Examination. By JULES SIMON. (*Le Progrès Méd.*, 1897, September 4, No. 36.)

JULES SIMON relates the following case:—Boy, aged 10, only child, admitted to Enfants-Malades in 1889 for pains in legs. Nothing hereditary of importance. Had pains in legs at 2½. dysentery at 5, after which he walked badly. In 1891, following was observed: strumous aspect, chronic blepharo-conjunctivitis, old keratitis, not intelligent-looking, speech slow, jerky. No nystagmus, club-foot, scoliosis, sensory troubles or sphincter affection. Knee-jerks absent. Standing difficult, especially if eyes shut. "Drunken" gait, often falls. Diagnosis of Friedreich's ataxia, confirmed by Joffroy. In 1896, slight tremor in head, jerky and fibrillary movements in hands; motion and sensation normal in upper extremities. Lower limbs cannot support him. Atrophy of calves and muscles of soles, club-foot, no knee-jerks, normal sensibility, cold feet. Pupils normal, transverse nystagmus on lateral fixation. Tends to slaver, especially on laughing, slight deviation of mouth to right. Cannot whistle now but could formerly. No scoliosis. The pains he has from time to time consist in intense heat and pricking in calf regions. Intelligence mediocre. Often has headaches followed by vomiting preceded by giddiness. Incontinence of urine frequently.

Death sudden and unexpected.

Post-mortem.—Normal naked eye appearance of cerebrum and cerebellum; weight of latter normal. No histological examination (unfortunately *Rep.*).

Examination of cord by Philippe (three months in Müller's fluid, Weigert-Pal stain).—I. Posterior columns show loss of myeline sheaths. (1) Sacral region, level of third sacral root: rootzones more affected than the zones of endogenous fibres, the short commissural fibres; myelinic reticulum of posterior horns very poor. (2) Lumbar region: rootzones specially affected, also the centrum ovale of Flechsig; cornu-commissural zone nearly intact; myelinic reticulum of posterior horns affected as in sacral region. (3) Dorsal region: decoloration of posterior columns at maximum in upper dorsal region; spec. the lumbo-sacral fibres, near the posterior fissure; the myelinic reticulum in Clarke's column scanty. (4) Cervical region (sixth root): Goll's column nearly completely decolorized; the postero-external column also well decolorized but less so than in dorsal region. (5) Bulbar region: fasciculi gracilis and cuneatus much decolorized, the former more, representing as it does Goll's column.

II. Posterior roots, loss of myeline everywhere, more so where the adjacent posterior columns are most affected.

III. Lateral columns, affected all along cord, but less so than posterior column; pyramidal tract and cerebellar tract affected; direct pyramidal tract in upper dorsal and cervical regions.

Where the nerve tubes have gone, their place is taken by a dense sclerosed neuroglia, arranged in places in whorls (both in posterior and lateral columns); connective tissue thickening of pia mater over posterior aspect; considerable diminution in number and volume of cells in posterior horn and at base of anterior horn; also of column of Clarke in lumbar and dorsal regions.

Conclusion.—The cord is the seat of a combined sclerosis affecting the posterior columns, the pyramidal bundle and the direct cerebellar tract. This corresponds fairly well with the lesions hitherto found in cases of Friedreich's disease. In addition to the combined sclerosis of the white matter, there is a lesion of the gray substance.

LESION OF THE SUPERIOR PARIETAL LOBULE.

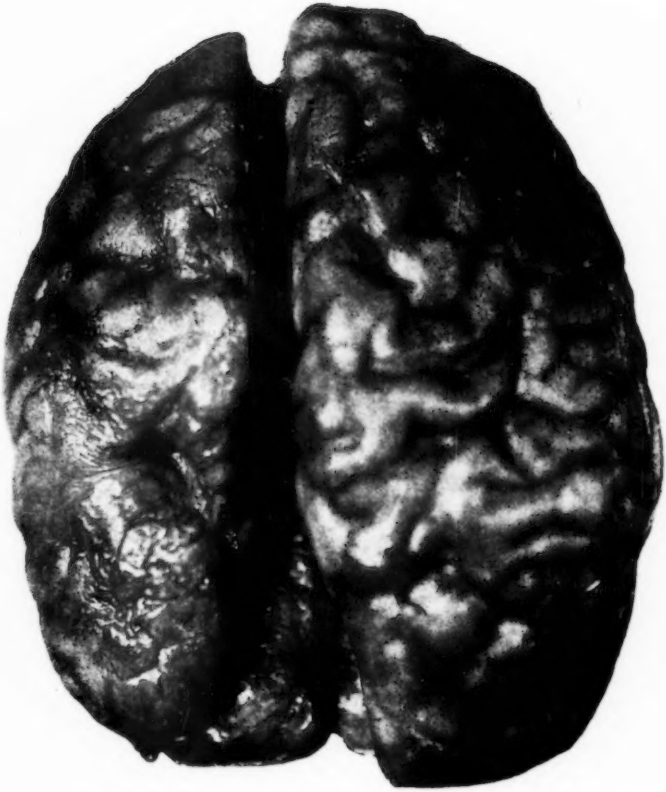
BY CECIL F. BEADLES.

ANY clinical evidence that may throw light on the areas of the cortical centres cannot but be of interest and importance. The following case has reference to the superior parietal lobule. A further point that makes this of some interest is the fact that definite lesions (except for congestion) of the brain of epileptics dying insane are but seldom seen.

The patient, 11445, was a youth who was admitted into Colney Hatch Asylum on November 25, 1891. He was then aged 18, and was the subject of epilepsy. His mother had died insane. The certificate stated that he was dull and of heavy expression, that he used bad and threatening language and had shown violence, and that he had been subject to fits since childhood. During the year 1892 he had frequent and severe fits, which occurred almost daily; he was weak-minded with impaired intelligence and almost incoherent, and was frequently violent in his behaviour. His health was fair and he was well nourished; was of clean habits but never occupied. In 1893 he continued much the same, although on the whole of quieter behaviour. Fits varied from several a week to three or four daily. They occurred both day and night. Some weakness of the right arm was noted.

In 1894 fits occurred daily. He was an impulsive and violent epileptic, continually quarrelling and fighting; his mental state was that of an imbecile. No change occurred the following year. Bromide of potassium was tried from time to time, but appeared to have but little effect on the epileptic paroxysms.

Up to January 6, 1896, the patient had two or three



Lesion of left superior parietal lobule. (*From a photograph.*)

fits almost daily. Then for twenty-four hours he had a large series of severe fits, more than thirty in number. The following day he had four more fits, and he died on January 8 after another succession of fits without regaining consciousness. Between the date of admission and death the patient had 3,360 fits (a rapid succession being counted as one); of this number 1,282 occurred during the last year of his life.

The special point of interest in the case is in reference to his right leg. The right ankle was drawn up and the toes were contracted. When he walked he dragged this foot and bore his weight on the left leg. No reference is made to this condition on the patient's admission, but it is probable that it then existed.

When he attempted to run, which almost invariably preceded a fit, his right foot seemed to trip him up. As he went off into the fit he would scream and roll about on the ground, crying out several times "Oh my leg, oh my leg." As he recovered from it the toes of the right foot were more contracted than usual.

At the autopsy the abdominal and thoracic viscera were normal with the exception of the heart, which was slightly hypertrophied with some excess of fat externally, and a small amount of atheroma above the aortic valves. It was found that the skull was greatly thickened throughout. The external conformation and measurements of the cranium were of fair size, but the internal capacity was distinctly small: all bony prominences within the fossæ were very marked. The dura mater was inherent to the calvaria and a considerable excess of fluid existed.

The whole brain was reduced in size, but the left cerebral hemisphere was much more contracted than the right. The left side was covered by a thickened milky pia arachnoid membrane, while the right was not thickened but was congested. The pons and medulla appeared smaller on the right side, and the right cerebellar lobe also seemed slightly reduced in size; the membrane over both these lobes was congested but not noticeably thickened. The weight of the whole brain before examination was 36½ ounces.

A localised lesion existed on the upper and hinder part of the parietal lobe of the left cerebral hemisphere. Situated in the hinder part of the superior parietal lobule, immediately in front of the parieto-occipital fissure, was a depression on the surface of the brain of the size of a fair sized marble. This was filled in the recent state by fluid in the meshes of the thickened pia mater. We have no evidence as to the cause of the lesion; no injury is known to have occurred. This lesion evidently involved the grey cortex. It is the site representing the area attributed to the centre for movement of the small toes, and this is fully borne out by the clinical evidence given above.

Gowers ("Diseases of the Nervous System," 1893, vol. ii., p. 17), in his explanation to a diagram illustrating the cortical centre for the leg, suggests that the centre does not extend as far back as the parieto-occipital fissure on the outer aspect of the hemisphere, but it will be seen in this case that the atrophy of the convolution reaches that point, and also extends down in front of that fissure on the inner surface of the hemisphere.

PROCEEDINGS OF THE NEUROLOGICAL SOCIETY OF LONDON.

At the Clinical Meeting of the Society, held at 11, Chandos Street, W., on Thursday, February 24th, the following cases were shown :—

Dr. DONKIN.—(1) A child, about 4 years old, with spasm of the extremities, more marked in the legs than in the arms, especially, though not entirely, occasioned by attempts at using them; associated with great coldness and blueness of feet. In some respects resembles a case of Thomsen's disease. The child was quite well and walked without difficulty until aged 2½ years. No history of any morbid heredity. There is no wasting or other abnormality discoverable on examination of the nervous, muscular, and other systems. (2) A man, aged about 50, with hemianæsthesia (including special senses) alternately appearing and disappearing on the same side, associated with weakness of the affected limbs. (The man is readily thrown into the hypnotic sleep.) The right field of vision is much contracted and the left almost *nil*. He protrudes his tongue towards the right, the left side of the tongue being markedly humped and the right angle of the mouth drawn downwards and to the right. When taste is being tested the tongue is protruded normally, and straight. There is probably a certain element of malingering in this case, but at first the symptoms were clearly genuine. (3) A case of paralysis agitans in a man aged 43. The tremors are only noticeable on use of the limbs. The motor symptoms and difficulty in walking were preceded by a long period of much aching pain in back and legs. The symptoms of propulsion and retropulsion are strongly marked, and the facial expression and bodily attitude are characteristic.

Dr. MOTT.—A woman, aged 50, married, seven children, two born dead at full term. Present illness commenced four years ago, immediately after the menopause, with numbness in the hands. The hands became swollen, and she had frequently pins and needles in them. March, 1897, numbness occurred in the chin,

followed by pins and needles, and the pain and numbness gradually spread to the lips and cheeks; the tongue also felt large, and the skin of the lower part of the face tense and swollen, with diminished sensation. Her hands and feet are swollen and somewhat cyanosed; they feel numb, and she does not feel well in the tips of the fingers. There is *diminished sensibility* to pain, pressure and thermal stimuli over the structures supplied by the two inferior divisions of the fifth nerves; the ophthalmic branch of the fifth is not affected. There is loss of taste over the anterior two-thirds of the tongue to salt, acid or sweet fluids; at the back of the tongue she tastes quite well. She did not smell assafœtida or ammonia. There was an erythematous rash over the upper part of the chest and neck.

Dr. HERBERT HAWKINS.—A lad, aged 18. About four years ago noticed that he had to lift his feet high in order to clear the ground in walking, and two years later noticed wasting of muscles of both thumbs. Nothing in family or personal history to throw light on this. Now, has great wasting and complete paralysis of muscles supplied by anterior tibial nerves on both sides, with slighter affection of peronei: great wasting of thenar and hypothenar muscles, with slight wasting and weakness of extensor muscles in forearms. No affection of any form of sensation. Fibrillar contractions in quadriceps extensor on both sides. Knee-jerks, present last November, disappeared in December. Well-marked nystagmus. Electrical reactions; true R.D. in left posterior inter-osseous group of muscles, and in anterior tibial groups on both sides; reduction of irritability to both currents in right posterior inter-osseous group, in intrinsic muscles of hands and in peronei.

Dr. BEEVOR.—Case of syphilitic meningitis. A man, aged 25. Syphilis and gonorrhœa twelve months previously. Onset in August, 1897, with malaise, severe headache, vomiting, followed in five days with severe pains in both arms and difficulty in moving them. No anæsthesia, no tingling, Glycosuria for a week. No pyrexia. At present the following muscles are paralysed on both sides:—Trapezius below level of second dorsal spine, exposing rhomboids which can be seen; serratus magnus; deltoids, except anterior and posterior parts, on the right. The following are weak:—Latissimus dorsi, pectoralis major, triceps. At rest the scapula was tilted, bringing the lower end nearer to the spinal column; no power to advance or abduct humerus beyond horizontal line, the latter being performed by supraspinatus acting without the deltoid.

Dr. Leonard Guthrie.—Unilateral syringo-myelia. Female, aged 29 years. Thermo-anæsthesia, analgesia of left side of head, neck and face, with unilateral seborrhœa capitis; nystagmus, paralysis of left side of palate and of left vocal cord. The bulbar symptoms are of two years' duration. They were formerly associated with analgesia and thermo-anæsthesia of the left upper extremity.

Dr. Stephen Mackenzie.—"A Case of Nodding Spasms (*Spasmus nutans*)." The patient is an infant, one year of age, who has not yet erupted any tooth. The spasms consist of combined nodding and rotatory movements of the head. As usual in such cases, these movements are combined with nystagmus, in this case in a lateral direction. The child is well nourished, and in other respects appears well.

Dr. Ormerod.—General athetosis, with double equino-varus, of uncertain origin, in a lad aged 19. Duration of disease five years.

Movements.—Slow, irregular wriggling movements of head, trunk and limbs, producing varied positions of the body, and occasionally throwing patient out of bed or chair. Larger joints more affected than small. Movements constant, except that they intermit temporarily when he uses limbs voluntarily, and that they cease during sleep; physiognomy peculiar; occasional slow grimacing; speech slovenly, nasal, sometimes almost unintelligible; breathing irregular and grunting.

Gait and condition of feet.— Double talipes equinus and pes cavus; dorsiflexion of metatarso-phalangeal joints of toes; contraction of calf-muscles; walks entirely upon his toes. Apparently intelligent, and has always been so; passed fifth standard at school.

History.—Present condition developed without assignable cause and gradually, at age of 14, after leaving school; movements said at first to have occurred only during voluntary actions. Father said to be nervous and alcoholic; family history otherwise negative.

Dr. F. E. Batten.—"Friedreich's Disease and Mental Defect." Boy, 11½ years old; eldest of family of seven; natural labour; late in learning to walk; late in learning to talk; always backward; unsteadiness in walking (increased during the last two years); dull boy; ataxia; speech defect (high palate); nystagmus (not constant); inco-ordination of arms; knee-jerks absent, pes cavus. Mother had epileptic fits before marriage. No hereditary history.

Dr. ALDREN TURNER.—A youth, aged 22, who had convulsions in infancy, but no subsequent fits. The present condition is as follows: — Complete diplopia facialis; convergent strabismus, owing to paralysis of both external recti muscles; double club-foot; a spastic gait and exaggeration of the knee-jerks; defect in the finer movements of the hands (one year ago there were irregular athetoid movements of the fingers); myopic astigmatism; no affection of sensation.

The Society held its first meeting out of London on Saturday, April 23rd, when, in response to an invitation from Dr. Alex. Hill, Vice-Chancellor of the University of Cambridge, a meeting was held in the Physiological Laboratory of that University, at 4.30 p.m., when the following communications were made:—

Dr. ALEX. HILL.—"Some Specimens of Granules."

Professor MACALISTER.—"The Race-Characters of Australian Brains."

Dr. LANGLEY.—"Some points relating to the Sympathetic System."

Dr. GASKELL (for Miss Alcock).—"The Distribution of the Facial Nerve in Ammocœtes."

Dr. F. E. BATTEN.—(1) "Nerve Terminations in Muscle Spindles." (2) (for Dr. Sihler, of Ohio).—"Motor Nerve Termination *within* the Muscle Spindle."

Dr. MOTT.—"Degeneration of the Neuron."

Dr. Langley's communication was an experimental demonstration, and the other papers were illustrated by lantern slides and by macroscopic and microscopic specimens.

At the conclusion of the meeting, members and visitors were entertained at dinner by the Vice-Chancellor, in Downing Hall.